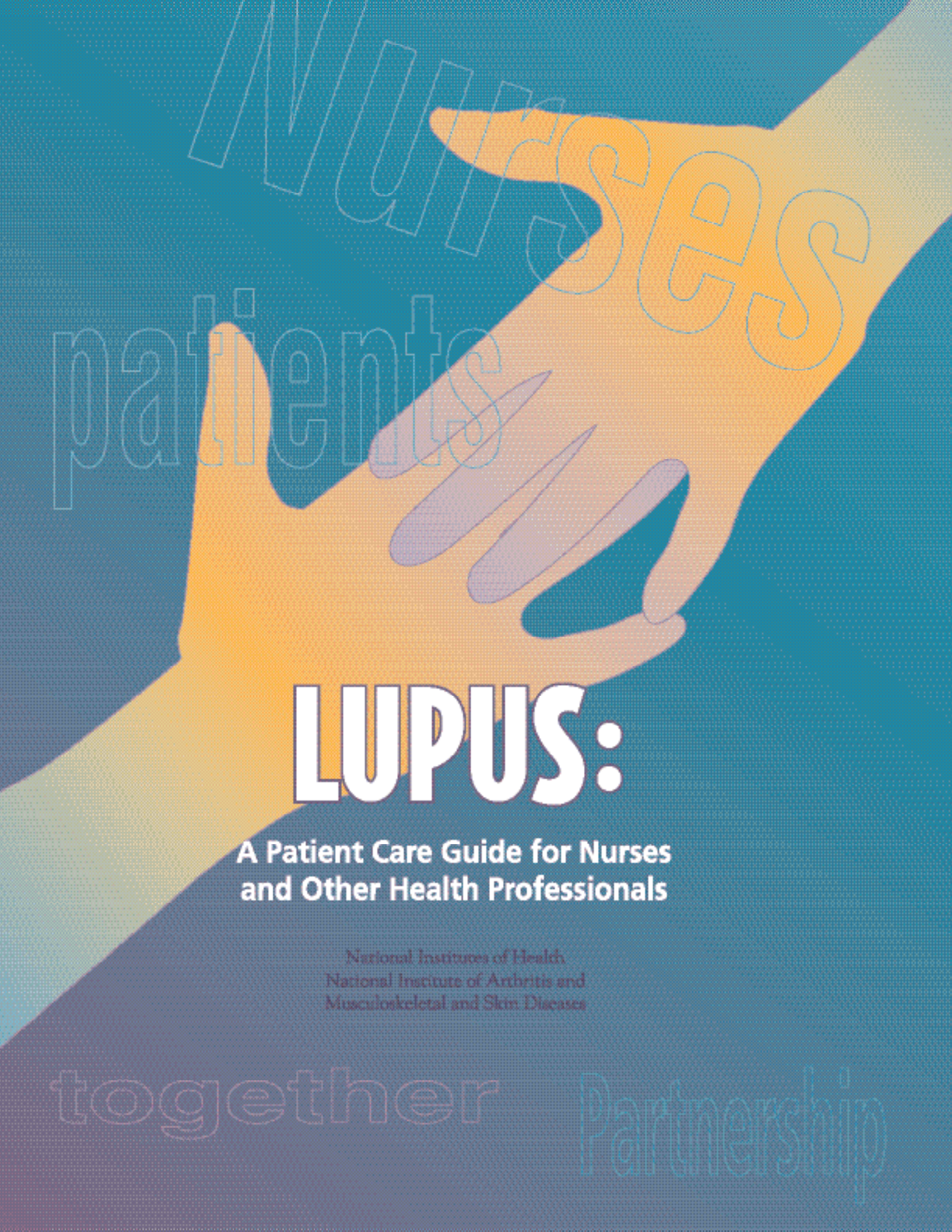

LUPUS:

A Patient Care Guide for Nurses and Other Health Professionals

National Institutes of Health
National Institute of Arthritis and
Musculoskeletal and Skin Diseases

For Your Information

This publication contains information about medications used to treat the health condition discussed in this booklet. When this booklet was printed, we included the most up-to-date (accurate) information available. Occasionally, new information on medication is released.

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LUPUS: A Patient Care Guide for Nurses and other Health Professionals is intended to provide an overview of lupus and how to care for patients who have the disease. It is not intended to provide medical guidelines for diagnosing and treating lupus, nor is it intended to be all inclusive. Specific medical advice is not provided, and NIAMS urges readers to consult with a qualified physician for diagnosis and for answers to individual questions.

LUPUS:

A Patient Care Guide for Nurses and Other Health Professionals

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LUPUS: A Patient Care Guide for Nurses and Other Health Professionals is an update of *Lupus Erythematosus: Handbook for Nurses* by Terri Nass, RN, which provided health care professionals with a comprehensive and detailed review of lupus. The National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS) is pleased to have had the opportunity to update the *Handbook* with the gracious permission of Ms. Nass.

Lupus erythematosus is a complex and challenging disease that affects the lives of many thousands of individuals and their families. It is multidimensional, with physical, emotional, and psychosocial aspects that require, in turn, multidimensional and patient-centered treatment and support strategies. This comprehensive guide brings together information on a wide variety of issues that health professionals need to know about in order to provide those treatment and support strategies for their patients with lupus. The guide covers general background on lupus, new advances in research, laboratory tests used to diagnose and evaluate lupus, care of the lupus patient, medications used to treat lupus, psychosocial aspects of lupus, and patient education and information.

A key element of the guide is that it contains information useful to the whole patient care team — nurses, physicians, physical and occupational therapists, social workers, and patients themselves.

Many people worked on this revision to incorporate the immense knowledge obtained over recent years on lupus and its management and to create a reader-friendly and useful book. We wish to thank all those who have played a role in bringing this guide to fruition, in particular the Task Force on Lupus in High Risk Populations, which was lead for a time by Lawrence E. Shulman, MD, PhD, the founding director of NIAMS, and which contributed much to ensuring that lupus remains in the public eye and on the research docket. In addition, we thank Constance Raab and Barbara Weldon in the NIAMS Office of Communications and Public Liason (OCPL) for supporting the update of this guide, and Ann Taubenheim of Cygnus Corporation for directing the project. Jacquelyn Jones-Gibson, RN, BSN, identified areas to update, and Julia Freeman, PhD, and Anne Brown Rodgers edited the manuscript. Phyllis Payne, MPH, wrote Chapter 2, *Advances in Lupus Research*, and Carolyne M. Albright, MS, RN,

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We hope that nurses and other health professionals across the country will find this new guide informative and useful as they work with people with lupus and their families.

INTRODUCTION

Lupus erythematosus is a unique, complex disease with a wide scope of symptoms. It is also an elusive condition in that it affects individuals differently and often does not follow a predictable course. For example, a patient who appears to be in remission during a routine office visit can present at an emergency room the following week with severe pericarditis or sudden stroke.

A person diagnosed with lupus may have symptoms and disease activity that are easily managed with treatment, but it is not uncommon for health care professionals to encounter a lupus patient with numerous severe symptoms that are difficult to control. No two lupus cases are alike. As a result, care of the patient with lupus erythematosus is a challenge that draws on all the resources, knowledge, and strengths the health care team has to offer. Each member of the health care team — physician, nurse, therapist, dietitian, social worker, and others — has an important role to play in treating specific aspects of the disease and in supporting the patient to cope with his or her condition.

LUPUS: A Patient Care Guide for Nurses and Other Health Professionals provides an overview of lupus erythematosus and the elements involved in caring for patients with this disease. It focuses on systemic lupus erythematosus (SLE). The *Lupus Guide* is intended primarily for nurses and other health professionals who work on an ongoing basis with lupus patients, and it provides the tools these professionals need to positively influence the care provided to this important group of patients. Many of these patients will be cared for in an outpatient setting, but the *Lupus Guide* also addresses the needs of hospitalized patients.

Each chapter of the *Lupus Guide* deals with a specific aspect of the disease. Chapter 1, *Lupus Erythematosus,* provides a general overview of the disease, including brief discussions of the diagnosis, treatment, medications, and psychosocial aspects. The chapter closes with a discussion of the implications of these issues for the ways that nurses and other health professionals provide care to lupus patients.

Chapter 2, *Advances in Lupus Research*, provides an overview of the current understanding of the etiology of lupus and describes areas of ongoing research. Recent research advances have significantly increased the understanding of lupus, and, as current research efforts unfold, there is hope for new treatments, improvements in patients' quality of life, and perhaps prevention of or cure for the disease.

Chapter 3, *Laboratory Tests Used to Diagnose and Evaluate SLE*, summarizes the main laboratory tests used to determine whether a patient has lupus and to monitor a patient's condition. These assessments include blood tests, measurements of autoimmunity, and tests for kidney disease.

Chapter 4, *Care of the Lupus Patient,* gives a system-by-system overview of the most common lupus manifestations and suggests nursing interventions. Several other key issues are also discussed, including general manifestations, pregnancy, infection, and nutrition. Not every patient will experience all of the problems and symptoms described. However, it is important for health professionals to be familiar with the range of possible manifestations so that they can accurately assess their patients and develop sound treatment and care plans.

Chapter 5, *Medications Used to Treat Lupus,* covers the major categories of drugs currently used to control lupus symptoms: nonsteroidal anti-inflammatory drugs, antimalarials, corticosteroids, and immunosuppressives. These medications can often be used successfully to treat lupus symptoms, but they are not without their own concerns. Chapter 5 discusses each category of medication, describes its mechanism of action and use in treating lupus, and reviews the potential side effects associated with it.

Chapter 6, *Psychosocial Aspects of Lupus,* provides information on this critically important aspect of the disease. Because of the chronic, unpredictable, and evolving nature of lupus, patients often have to cope with serious emotional and psychosocial issues along with the physical dimensions of their illness. A good understanding of these issues will help nurses and other health professionals provide the empathetic and supportive care lupus patients need.

Chapter 7, *Patient Information,* contains 16 short fact sheets covering a broad range of issues related to lupus. Five of the fact sheets deal with lupus medications. These Patient Information Sheets are designed to help patients understand their disease and its symptoms and complications and to

develop effective ways of living with and controlling lupus. Nurses and other health professionals will find the sheets helpful in their ongoing teaching efforts.

The *Lupus Guide* closes with a chapter of resources for further information on lupus and a bibliography of the source materials used to develop the book. The bibliography has been updated for the 2001 edition of the guide. Furthermore, an addendum has been added that refers readers to a source of information on new drugs.

Today, the prognosis for people with lupus is far brighter than it was even 20 years ago. Advances in research, improved treatments, a growing list of support networks and information resources, and an increased emphasis on close cooperation between the patient and her or his health care team mean that, for many patients, it is possible to have lupus yet remain active and involved with life, family, and work. The *Lupus Guide* is intended to provide nurses and other health professionals with a solid grounding in this important disease so that they can provide the care to make an active and involved life a reality for women and men with lupus.

Chapter One

LUPUS ERYTHEMATOSUS

Care for the patient with lupus erythematosus is a challenge that draws on all the resources, knowledge, and strengths the health care team has to offer. Because of the unpredictable, highly individualized, and frequently changing nature of the disease as well as the intricacy of each patient's needs, it is impossible to predict the treatment for one patient from the outcome of treatment for another. Careful listening to the person's concerns; a cooperative, multidisciplinary approach; and a flexible plan of care will provide the patient with consistent, supportive care and the reassurance that her or his needs are being attended to.

DEFINITION AND DESCRIPTION

Lupus means "wolf." **Erythematosus** means "redness." In 1851, doctors coined this name for the disease because they thought the facial rash that frequently accompanies lupus looked like the bite of a wolf. Lupus can be categorized into three groups: discoid lupus erythematosus, systemic lupus erythematosus, and drug-induced systemic lupus erythematosus.

Discoid lupus erythematosus (DLE) is characterized by a skin rash only. It occurs in about 20% of patients with systemic lupus erythematosus. The lesions are patchy, crusty, sharply defined skin plaques that may scar. These lesions are usually seen on the face or other sun-exposed areas. DLE may cause patchy, bald areas on the scalp and hypopigmentation or hyperpigmentation in older lesions. Biopsy of a lesion will usually confirm the diagnosis. Topical and intralesional corticosteroids are usually effective for localized lesions; antimalarial drugs may be needed for more generalized lesions. DLE only rarely progresses to systemic lupus erythematosus.

Systemic lupus erythematosus (SLE, or lupus) is a chronic, inflammatory, multisystem disorder of the immune system. *LUPUS: A Patient Care Guide for Nurses and Other Health Professionals* is concerned primarily with this form of lupus. In SLE, the body develops antibodies that react against the person's own normal tissue. This abnormal response leads to the many manifestations of SLE and can be very damaging. The course is unpredictable and individualized; no two patients are alike. Lupus is not contagious, infectious, or malignant. It usually develops in young women of childbearing years, but many men and children also develop lupus. African Americans and Hispanics have a higher frequency of this disease than do Caucasians. SLE also appears in the first-degree relatives of lupus patients more often than it does in the general population, which indicates a strong hereditary component. However, most cases of SLE occur sporadically, indicating that both genetic and environmental factors play a role in the development of the disease.

Lupus varies greatly in severity, from mild cases requiring minimal intervention to those in which significant and potentially fatal damage occurs to vital organs such as the lungs, heart, kidney, and brain. The disease is characterized by "flares" of activity interspersed with periods of improvement or remission. A flare, or exacerbation, is increased activity of the disease process with

DRUGS IMPLICATED AS ACTIVATORS OF SLE

Drugs with proven association

- Chlorpromazine
- Hydralazine
- Isoniazid
- Methyldopa
- Procainamide

Drugs with possible association

- Beta blockers (e.g., acebutolol, atenolol, labetalol, metoprotolol, oxprenolol, pindolol, practolol, and propranolol)
- Captopril
- Carbamazine
- Cimetidine
- Diphenylhydantoin (phenytoin)
- Ethosuximide
- Methimazole
- Penicillamine
- Phenazine
- Quinidine

an increase in physical manifestations and/or abnormal laboratory test values. Periods of improvement may last weeks, months, or even years. The disease tends to remit over time. Some patients never develop severe complications, and the outlook is improving for those patients who do develop severe manifestations.

Drug-induced SLE develops after the use of certain drugs and has symptoms similar to those of SLE. The characteristics of this syndrome are pleuropericardial inflammation, fever, rash, and arthritis. Serologic changes also occur. The clinical and serologic signs usually subside gradually after the offending drug is discontinued. A wide variety of drugs is implicated in this form of SLE (see box, opposite).

SYMPTOMS OF SLE

Early symptoms of SLE are usually vague, nonspecific, and easily confused with other pathological and functional disorders. Symptoms may be transient or prolonged, and individual symptoms often appear independently of the others. Moreover, a patient may have severe symptoms with few abnormal laboratory test results, and vice versa. The box on the next page lists the range of clinical symptoms seen in patients with lupus over the lifetime of the disease.

DIAGNOSIS OF SLE

The onset of lupus may be acute, resembling an infectious process, or it may be a progression of vague symptoms over several years. As a result, diagnosing SLE is often a

challenge. A consistent, thorough medical examination by a doctor familiar with lupus is essential to an accurate diagnosis. This must include a complete medical history and physical examination, laboratory tests, and a period of observation (possibly years). The doctor, nurse, or other health professional assessing a patient for lupus must keep an open mind about the varied and seemingly unrelated symptoms that the patient may describe. For example, a careful medical history may show that sun exposure, use of certain drugs, viral disease, stress, or pregnancy aggravates symptoms, providing a vital diagnostic clue.

SYMPTOMS OF SLE

- Arthralgia
- Arthritis
- Fever (>100 °F)
- Skin rashes
- Anemia
- Kidney damage
- Pleurisy
- Facial rash
- Photosensitivity
- Alopecia (hair loss)
- Raynaud's phenomenon
- Seizures
- Mouth or nose ulcers

No single laboratory test can definitely prove or disprove SLE. Initial screening includes a complete blood count (CBC), liver and kidney screening panels, laboratory tests for specific autoantibodies (e.g., antinuclear antibodies [ANA]), a syphilis test (VDRL), urinalysis, blood chemistries, and erythrocyte sedimentation rate (ESR). Abnormalities in these test results will guide further evaluations. High-titer anti-nDNA antibody or anti-Sm antibody are important indications of lupus. Specific immunologic studies, such as those of complement components (e.g., C3 and C4) and other autoantibodies (e.g., anti-La and anti-Ro), are used to help evaluate the patient's immune status and to monitor the activity of the disease. At times, biopsies of the skin or kidney using immunofluorescent staining techniques can support a diagnosis of SLE (see Chapter 3, *Laboratory Tests Used to Diagnose and Evaluate SLE*, for further information). A variety of laboratory tests, X rays, and other diagnostic tools are used to rule out other pathologic conditions and to determine the involvement of specific organs. It is important to note, however, that any single test may not be sensitive enough to reflect the intensity of the patient's symptoms or the extent of the disease's manifestations.

The American College of Rheumatology (ACR), an organization of doctors and associated health professionals who specialize

in arthritis and related diseases of the bones, joints, and muscles, has developed and refined a set of diagnostic criteria (see box, opposite). If at least 4 of the 11 criteria develop at one time or individually over any period of observation, then the patient is likely to have SLE. However, a diagnosis of SLE can be made in a patient having fewer than four of these symptoms.

ACR CRITERIA FOR DIAGNOSING SLE

- Malar rash
- Discoid rash
- Photosensitivity
- Oral ulcers
- Arthritis
- Serositis (pleuritis or pericarditis)
- Renal disorder (persistent proteinuria or cellular casts)
- Neurological disorder (seizures or psychosis)
- Hematologic disorder (anemia, leukopenia or lymphopenia on two or more occasions, thrombocytopenia)
- Immunologic disorder (positive LE cell preparation, abnormal anti-DNA or anti-Sm values, false-positive VDRL syphilis test)
- Abnormal ANA titer

Source: Tan E. The 1982 required criteria for the classification of systemic lupus erythematosus. *Arthritis and Rheumatism* 1982;25:1271–1277.

TREATMENT OF SLE

The treatment of SLE is as varied as its course. Although there is no cure for lupus and it is difficult to predict which treatment will be most effective for each patient, there have been significant gains in treating patients, and there is general consensus on several treatments.

A conservative regimen of physical and emotional rest, protection from direct sunlight, a healthful diet, prompt treatment of infections, and avoidance of known allergens and aggravating factors are the mainstays of lupus therapy. In addition, for female patients, pregnancy must be planned for times when the disease is in remission.

Physical Rest

This basic component of everyone's good health is essential for the lupus patient. The fatigue of lupus is not sleepiness or tiredness from physical exertion, but rather a frequent, persistent complaint often described as a "bone-tired feeling" or a "paralyzing fatigue." Normal rest often does not refresh the patient or eliminate the tiredness due to lupus, and fatigue may persist despite normal laboratory test results. The patient and family need instruction on how to use this tiredness as a guide to activity and when the person should stop for rest. It must be

reinforced that this need for rest is not laziness. Restful sleep of 8–10 hours per night, naps, and "timeouts" during the day are basic guidelines; strict bed rest is usually not required. Physical activity should be encouraged as the patient can tolerate it. An individualized exercise routine may facilitate recovery from a flare and promote well-being.

Emotional Rest

A patient's emotional stressors should be carefully assessed, because they may play a role in triggering a flare. The patient should be instructed on how to avoid these stressful situations. However, the physical manifestations of lupus must be treated as they present themselves while the emotional stresses are explored. Discussions with the family on this issue are essential for providing information and in obtaining their support. Counseling for both the patient and the family may be an option. Chapter 6, *Psychosocial Aspects of Lupus*, explores these issues in further detail.

Protection From Direct Sunlight

Photosensitivity is an abnormal reaction to the ultraviolet (UV) rays of the sun and results in the development or exacerbation of a rash that is sometimes accompanied by systemic symptoms. About one-third of lupus patients are photosensitive. All lupus patients should avoid direct, prolonged exposure to the sun. Sun-sensitive patients should frequently apply a sunscreen with a Sun Protection Factor (SPF) of at least 15, avoid unprotected exposure between 10 a.m. and 4 p.m., and wear protective clothing, such as wide-brimmed hats and long sleeves. Lupus patients should be aware that UV rays are reflected off water and snow, and that glass, such as car windows, does not provide total protection from UV rays.

Lupus patients should also know that fluorescent and halogen lights may emit UV rays and can aggravate lupus. This may be an issue for patients who work in offices lit by these kinds of lights. Sunscreen and protective clothing can help minimize exposure, and plastic devices are available that block UV emissions from fluorescent or halogen light bulbs.

Diet and Nutrition

A well-balanced diet is essential in maintaining good health for all people, including lupus patients. There are currently no specific dietary recommendations or limitations for those with lupus, but a restricted diet plan may be prescribed when fluid retention, hypertension, kidney disease, or other problems are present. Food intolerances and allergies may occur, but there is no evidence that these are more common in lupus patients than in the general population.

The health professional should make a note of the patient's dietary history and suggest diet counseling if appropriate, especially if the patient has a problem with weight gain, weight loss, gastrointestinal (GI) distress, or food intolerances. Nutritional considerations in treating lupus patients are discussed further in Chapter 4, *Care of the Lupus Patient*.

Treatment of Infections

Prompt recognition and treatment of infection is essential for those with lupus. However, cardinal signs of infection may be masked because of SLE treatments. For example, a fever may be suppressed because anti-inflammatory therapy is being given. When an infection is being treated, the health professional should be alert to medication reactions, especially to antibiotics.

Pregnancy and Contraception

Spontaneous abortion and premature delivery are more common for women with SLE than for healthy women. To minimize risks to both mother and baby, a pregnant woman with lupus should be closely supervised by an obstetrician familiar with lupus. The safety of oral contraceptives for women with lupus is currently under investigation. The use of an intrauterine device (IUD) is not recommended because of the lupus patient's increased risk of infection.

Surgery

Surgery may exacerbate the symptoms of SLE. Hospitalization may be required for otherwise minor procedures, and postoperative discharge may be delayed. If it is elective, the surgery (including dental surgery and tooth extraction) should be postponed until lupus activity subsides.

Immunizations

Immunizations with killed vaccines have not been shown to exacerbate SLE. However, live vaccines with attenuated organisms are not advisable. A lupus patient should consult her or his doctor before receiving any immunizations, even routine ones.

MEDICATIONS FOR SLE

SLE management should include as few medications for as short a time as possible. Some patients never require medications, and others take them only as needed or for short intervals, but many require constant therapy with variable doses. Despite their usefulness, no drugs are without risks. Medications frequently used to control the symptoms are nonsteroidal anti-inflammatory drugs (NSAIDs), antimalarials, corticosteroids, and immunosuppressives. Other medications may be necessary to control specific manifestations. Before prescribing a medication, it is

helpful to scrutinize a patient's past response to treatments. A careful drug history should be taken; in particular, hypersensitivities or allergies to certain drugs should be noted, as these may aggravate the lupus. Patient and family education about medications and their side effects is essential. Chapter 5, *Medications Used to Treat Lupus*, presents more detailed information on this issue, and Chapter 7, *Patient Information*, includes relevant information for patients.

PSYCHOSOCIAL ASPECTS

For the lupus patient, the emotional aspects of dealing with a chronic disease can be overwhelming. They can also make a patient feel isolated from friends, family, and coworkers. Grief, depression, and anger are common reactions of patients about their lupus.

Lupus patients and their families deal with the disease in strikingly different ways. Managing the ups and downs of the disease may put strains on relationships and marriages. Younger patients may fail to assert their independence or develop a life away from home if they feel they cannot cope with their disease on their own. Family members are often confused and frightened over the changes they see and need guidance and constructive suggestions on helping the patient. Children of lupus patients, particularly those too young to really understand the disease, may need special help in coping with their parent's illness. It is in these areas that the patient, family, and support systems need to be assessed, encouraged, and guided so that they work together as a team. Allowing the patient and her or his family time and freedom to move through different emotional phases without criticism and unrealistic expectations will facilitate acceptance of the disease. The health professional can have a major role in helping a patient adjust and can help with referrals to a social worker, counselor, or community resource, if needed. Chapter 6, *Psychosocial Aspects of Lupus*, discusses these issues in more detail.

HEALTH CARE IMPLICATIONS

How lupus is defined, diagnosed, and treated and the psychosocial issues involved have implications for the way that the nurse or other health professional works with a patient who has lupus. For example, a newly diagnosed lupus patient needs help in getting current, accurate information about the disease and in defining realistic expectations and goals. The Patient Information Sheets in Chapter 7 can help. The health professional can clarify information with the patient's doctor, make rounds with the doctor, and act

as a liaison between the patient and the doctor, if needed. Frequently, many doctors are involved in caring for a lupus patient at one time. This may increase the patient's confusion and leave gaps in information. Emotional support to the patient is essential. Being available for questions, providing reassurance, and encouraging discussion of fears and anxieties are all crucial roles that the nurse can play.

The lupus patient hospitalized during a flare requires symptomatic nursing care. It is important to note that objective data, such as anemia or sedimentation rate, may not support subjective complaints of fatigue or pain. Careful head-to-toe assessment and documentation of all symptoms and complaints are important. Symptomatology changes constantly, so frequent reassessment is necessary. Reevaluations validate a patient's concerns and alert the doctor to changes that may be transient yet significant.

The patient's tolerance for physical activity and need to control what she or he can do should be respected. The patient should be involved in developing a care plan and daily schedule of activities.

The best way to treat lupus is to *listen to the patient*, whether that patient was diagnosed today or years ago. The patient's support systems can be expanded to include pamphlets and books, physical or occupational therapy, vocational rehabilitation, homemaker services, the Visiting Nurses Association (VNA), the Lupus Foundation of America (LFA), the SLE Foundation, and the Arthritis Foundation(AF).

Lupus is a challenge to everyone concerned. The health professional has a key role in its management. Accurate documentation, supportive care, emotional support, patient education, and access to community resources will provide the patient and her or his family with the tools they need to cope effectively.

Chapter Two

ADVANCES IN LUPUS RESEARCH

Scientists know that SLE is an autoimmune disease, and recent studies have begun to unravel some of the elements involved. There is support for genetic, environmental, and hormonal components in its etiology. Much research is being conducted to understand these factors and how they work together.

Epidemiologic studies may yield further clues about the cause of lupus. For example, SLE is more prevalent in women, especially those in the reproductive years, than in men. And, while people of all races get lupus, the incidence rate for African-American women is three times higher than it is for Caucasian women. African-American women also tend to develop the disease at a younger age, develop more serious complications, and have a higher mortality rate from the disease than do Caucasian women. Researchers are trying to find out why lupus is more common in these populations.

Health professionals continue to search for better ways to care for lupus patients. Answers to what causes the disease and why certain people are more likely to develop it may one day lead to promising new treatments for or even prevention of the disease. In the meantime, researchers continue to look for new treatments and ways to modify existing ones to diminish or eliminate side effects and to improve quality of life for people who have lupus.

During the last decade, there has been a tremendous amount of progress in lupus research. The number of studies on this disease has increased exponentially, and most researchers believe that answers to some of the key questions are close at hand. This chapter highlights some of the recent research advances in lupus and provides an overview of the direction of current research.

ETIOLOGY

Investigators have found evidence to support several likely possibilities in the etiology of SLE. Some believe there may be more than one type of SLE and that its etiology may vary from one person to the next. Current studies are focusing on the following elements:

- immune system dysfunction
- genetics
- environmental influences
- hormones

In lupus research, as in many areas of research, animal models have played an important role. This discussion of the etiology of lupus includes examples of research conducted in animal models that illustrate how these factors might influence the development of SLE in humans.

Immune System Dysfunction

Lupus is an autoimmune disease, so called because a person's immune system attacks her or his own tissues. In lupus, the signs and symptoms of the disease can be attributed to damage caused directly by antibodies, the deposition of immune complexes (the combination of antigen and antibody), or cell-mediated immune mechanisms. A number of steps are involved in these mechanisms, and scientists hope to reveal the cause of lupus by examining each step. In the process of doing so, they also may find new ways to treat lupus.

One of the hallmarks of lupus is the formation of autoantibodies, which are antibodies that react with a person's own tissue. Autoantibodies occasionally can be present in healthy people, but they are typically found in low concentrations. Essentially all patients with lupus have autoantibodies, generally in high concentrations. The autoantibodies found in lupus patients are often called antinuclear antibodies because they generally target the nucleic acids, proteins, and ribonucleoprotein complexes inside a cell's nucleus. Other autoantibodies in lupus patients also can bind to cell surface membranes and destroy cells directly.

Research studies have shown an association between the presence of certain autoantibodies and particular manifestations of lupus, such as kidney or skin disease. Scientists are now trying to establish whether these autoantibodies actually cause signs or symptoms

of lupus. However, most people with lupus test positive for many different autoantibodies, so it is often very difficult to identify which autoantibodies are responsible for a specific type of tissue damage in human subjects.

In lupus, the immune system produces too many autoantibodies and forms too many immune complexes. Normally, antigen-antibody immune complexes are joined by complement, a substance in the blood that aids in the breakup and removal of immune complexes from the body. Scientists have found that SLE patients have both inherited and acquired abnormalities in complement and complement receptors. These deficiencies in complement may decrease the body's ability to get rid of immune complexes. Immune complexes not broken up may be deposited in various body tissues, leading to the inflammation that results in tissue damage. Scientists continue to study

- the nature of immune complexes and what happens to them once they are formed,
- the nature of the autoantibodies that make up the immune complexes, and
- the reason for increased production of autoantibodies.

Genetics

There is considerable evidence showing that genes play a role in the etiology of lupus. The extremely high occurrence of lupus in identical twins and the increased prevalence of lupus among first- and second-degree relatives of lupus patients suggests a genetic component. In addition, when researchers look at autoantibodies typically found in a lupus patient and her or his siblings and compare them with clinical manifestations of the disease in the individuals, they find that the individuals have the autoantibodies in common more often than they have the clinical manifestations in common. This finding indicates a genetic basis for the formation of autoantibodies that play a role in lupus.

Studies to date suggest that many different genes contribute to lupus susceptibility and that no single genetic abnormality causes the disease. It also appears that genes may be influential in determining the type or severity of lupus. For example, among African Americans with lupus, those with lupus nephritis are more likely than those with other clinical manifestations to have the gene for a form of a receptor that has a low efficiency for capturing immune complexes.

Other genes that have been associated with lupus in humans include

- the immune system genes human leukocyte antigen (HLA)-DR3 (and B8 in older data), HLA-DR2, and complement C4 genes;
- alleles at HLA-DR and HLA-DQ, which are associated with certain autoimmune characteristics found in lupus; and
- a polymorphism of the T-cell receptor, which has been associated with anti-Ro, one of the autoantibodies commonly found in mothers of babies with neonatal lupus erythematosus.

Researchers studying lupus in animals have recently discovered a single gene that causes a lupus-like illness in mice. In these mice, the *fas* gene, one of the genes that controls apoptosis (programmed cell death), is defective. When the defective *fas* gene is replaced with a normal gene, the mice no longer develop signs of the disease.

Scientists continue to study the genetics of lupus in humans and in animals. If the genes that create a genetic predisposition for lupus can be identified, it may be feasible to correct genetic defects through gene therapy or other treatments. At this time, researchers are studying

- genes associated with the clearance of immune complexes,
- genetic markers associated with immune abnormalities in lupus, and
- genes associated with apoptosis in lupus.

GLOSSARY OF IMMUNOLOGIC AND GENETIC TERMS

Allele—one of the two or more forms of a gene

Complement receptors—molecules on the surface of cells that react with complement

Intranuclear nucleic acids—deoxyribonucleic acid (DNA) and ribonucleic acid (RNA) found inside the nucleus of the cell

Polymorphism—a genetic characteristic that can be physically manifested in more than one form

Ribonucleoprotein complexes—molecules containing ribonucleic acids (RNA) and protein

Environmental Influences

Researchers believe that genetic predisposition is just one piece of the puzzle of lupus etiology. Studies have shown that the occurrence of lupus is high among both members of a pair of identical twins and much lower among nonidentical twins and other full siblings. The fact that this concordance is not 100% among identical twins, however, suggests that environmental agents probably trigger lupus in individuals with a genetic predisposition. Environmental factors that scientists are considering include sunlight,

stress, certain chemical substances, and infectious agents such as viruses.

Sunlight

Exposure to the UV rays of sunlight can lead to a skin rash and exacerbate systemic manifestations of lupus. Exposure to UVB light causes certain cellular proteins to accumulate in abnormally large amounts on the cell's surface. These proteins react with autoantibodies commonly found in people with SLE, leading to a local or systemic inflammatory response.

Stress

Doctors suspect stress is a possible trigger for lupus flares. Frequently, patients ascribe their first symptoms or worsening symptoms to a stressful event, such as divorce, death of a loved one, or job loss. Scientists do not have a clear explanation for this phenomenon, but research is being done to find out whether stress hormones such as adrenaline or cortisone may influence the development or course of the disease.

Chemical Substances

A number of drugs cause a lupus-like illness in susceptible individuals, including chlorpromazine, hydralazine, isoniazid, methyldopa, and procainamide. When the offending drug is stopped, the lupus symptoms resolve. When researchers determine how these drugs cause lupus, they may be able to provide further answers on the etiology of SLE.

Viruses

Many researchers suspect that infectious agents, such as viruses, may trigger lupus, somehow disrupting cellular immune function in susceptible individuals. It is possible that the virus infects B cells (cells programmed to produce antibodies in response to specific antigens) and causes them to produce autoantibodies. Researchers are studying various mechanisms by which viruses could result in autoimmunity.

Hormones

SLE is more prevalent in women during their reproductive years. In addition, disease activity sometimes flares during pregnancy or during the postpartum period. For these reasons, researchers have long considered that hormones may influence lupus. Some research in animals also supports this supposition. Lupus-like illnesses in animals are exacerbated when they receive female hormones. Studies are under way to find out more about how hormones may influence the course and development of lupus.

Improving current treatments for lupus patients and improving the reproductive health of women with lupus are also important elements of ongoing lupus research. Specifically, investigators are studying ways to

- minimize the use of immunosuppressives, such as corticosteroids and cyclophosphamide, to decrease unwanted side effects and improve the quality of life for lupus patients;
- develop new therapies with fewer side effects;
- correct underlying immune abnormalities; and
- improve women's reproductive health and evaluate the safety of hormone replacement therapy for women with lupus.

Minimize the Use of Immunosuppressives

Corticosteroids, such as prednisone, are a mainstay of lupus therapy because they suppress the immune system and reduce inflammation. Unfortunately, they also cause some serious side effects, including osteonecrosis, osteoporosis, and coronary artery disease. Other, less serious side effects can also take a toll on the patient's quality of life. Scientists are investigating how corticosteroid use can be minimized in such a way that their benefits are retained while their sides effects are reduced.

Cyclophosphamide also suppresses the immune system and has anti-inflammatory properties. Treatment with cyclophosphamide improves many severe manifestations of lupus. Unfortunately, cyclophosphamide can be toxic. Patients using this drug may experience gastrointestinal complications, alopecia, and an increased risk for infections. In the long term, cyclophosphamide also may damage gonadal tissue and lead to ovarian or testicular failure. Other potential long-term complications include hemorrhagic cystitis, bladder fibrosis, and bladder cancer. At this time, scientists are conducting studies to better understand the long-term effects of cyclophosphamide therapy. In addition, they are exploring the use of additional drugs that might counteract some of the negative side effects of cyclophosphamide, and trying to find the most effective dose regimen that causes the fewest severe side effects.

Scientists are also trying to identify combination therapies that may be more effective than single-treatment approaches. For example, in lupus nephritis patients with moderate kidney scarring, a combination of cyclophosphamide and prednisone is more

effective in preserving renal function than is treatment with prednisone alone. In these patients, the combination therapy reduces the likelihood of end-stage renal failure.

Develop New Therapies

While some researchers are examining existing drug and treatment practices, other researchers are developing new treatment regimens. Promising areas of treatment research include biologic agents, hormones, newer forms of chemotherapy, and nitric oxide.

Biologic Agents

On the basis of new information about the SLE disease process, scientists are using novel biologic agents to selectively block parts of the immune system. Developing and testing these new drugs, which are based on compounds that occur naturally in the body, is an exciting and promising new area of lupus research. Scientists hope that these naturally occurring substances will cause few side effects. In addition, use of these agents may yield clues to the etiology of the disease.

Hormones

Because hormones are believed to influence the course and perhaps even the etiology of lupus, many researchers are interested in testing the effects of hormones on lupus patients. For example, animal and human studies have shown benefits associated with dehydroepiandrosterone (DHEA) therapy. DHEA is a naturally occurring hormone present in unusually low concentrations in people with lupus. DHEA is currently being tested in clinical trials to determine if its use can improve the clinical outcome and reduce the prednisone requirements of lupus patients.

Chemotherapy

Researchers also continue to look for new forms of chemotherapy that work selectively on the immune system. For example, they are testing immunosuppressive drugs, such as cyclosporine and 2 chlorodeoxyadenosine, which strongly suppress immune function. Preliminary clinical studies suggest that these drugs may be used in treating patients with lupus nephritis.

Nitric Oxide

Recent studies sponsored by the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS) have investigated the role of nitric oxide, a natural substance known to promote inflammation. These studies, using mice that develop a lupus-like autoimmune illness, including joint and kidney inflammation, showed that the animals produce abnormally high levels of nitric oxide. When the mice were treated

with a drug that blocks nitric oxide formation, development of kidney disease was prevented and joint inflammation was reduced. Additional studies are needed to determine whether nitric oxide plays a role in inflammatory disease in humans and whether drugs that block the formation or action of nitric oxide will be valuable in treating patients with lupus.

Correct Underlying Immune Abnormalities

Researchers predict that one day it may be possible to correct the underlying immune abnormalities in people with lupus. Studies are under way to explore the dimensions, risks, and benefits of reconstructing the immune system by bone marrow transplantation and of using gene therapy to treat lupus.

Improve Reproductive Health in Women With Lupus

Because of recent improvements in diagnostic tools for lupus and a better understanding of the disease, doctors can now predict the likelihood of a lupus-related miscarriage and identify women at risk for giving birth to babies with neonatal lupus. Doctors and lupus patients can now take measures to prevent miscarriages, and doctors can prepare to treat those babies born with neonatal heart block, the most serious complication of neonatal lupus.

Progress is also being made in another important area of reproductive health. In the past, women with lupus have not been able to use oral contraceptives or take advantage of hormone replacement therapy because of concerns that estrogens exacerbate lupus. However, recent data suggest these drugs may be safe for some women with lupus, and a current study funded by NIAMS, the NIH Office of Research on Women's Health, and the NIH Office of Research on Minority Health is focusing on the safety and effectiveness of oral contraceptives and hormone replacement therapy in women with lupus. This clinical trial is called the Safety of Estrogen in Lupus Erythematosus National Assessment (SELENA). Researchers hope this study will yield options for safe, effective methods of contraception for young women with lupus as well as options for estrogen replacement therapy for postmenopausal women with lupus.

ROLE OF THE NATIONAL INSTITUTE OF ARTHRITIS AND MUSCULOSKELETAL AND SKIN DISEASES (NIAMS)

NIAMS leads and coordinates the Federal biomedical research effort in lupus by conducting and supporting research projects, research training, clinical trials, and epidemiologic studies and by disseminating information on research results.

NIAMS funds many scientists across the United States who are studying the causes and mechanisms of tissue injury in SLE and why lupus strikes women and certain minority populations more frequently. In addition, NIAMS has established the first Specialized Centers of Research (SCORs) devoted to lupus. These centers enable basic scientists and clinicians to collaborate closely on lupus research.

To further the study of the genetics of lupus and to provide a resource for all researchers in this field, NIAMS has established a lupus registry and repository. Researchers who study families in which two or more members have been diagnosed with lupus collect and continually update clinical, demographic, and laboratory data on these individuals and submit the data to the lupus registry. Blood, cell, and tissue samples and DNA from these individuals will be stored in the lupus repository.

The registry and repository will allow all lupus researchers access to an enormously valuable database of information on lupus patients. For example, researchers will be able to analyze each DNA sample in the repository for the presence of a standard set of genetic markers. A centralized database will maintain this genetic information along with clinical and laboratory information from the registry. Together, these data can be used as the starting point for genetic analysis to identify possible lupus genes. Finding the genes that cause the disease may help researchers develop new treatments. In addition, this research will help identify which lupus patients will develop the most severe manifestations of the disease. This will help doctors decide who needs the most aggressive treatment.

The research advances of the past have led to significant improvements in the prognosis for patients with lupus. As current research efforts unfold, there is continued hope for new treatments, further improvements in patient quality of life, and ultimately, for ways in which to prevent or cure the disease.

Chapter Three

LABORATORY TESTS USED TO DIAGNOSE AND EVALUATE SLE

Lupus is characterized by abnormalities in many laboratory test results. These abnormalities are different for every patient and vary significantly during the course of a patient's disease. The serial evaluation of an individual's tests along with the physician's observations and the patient's history determine the diagnosis of SLE, its course, and the treatment regimen. All laboratory values must be interpreted in light of the patient's present status, other correlating laboratory test results, and coexisting illnesses. This chapter briefly describes the major tests used to diagnose and evaluate SLE and provides information on their rationale and clinical usefulness. Nurses and other health professionals should consult rheumatologists, manuals of laboratory and diagnostic tests, or hospital clinical laboratory departments for further information on possible interpretations of results from these tests and their implications for SLE.

TESTS FOR BLOOD CELL ABNORMALITIES

Blood cell abnormalities often accompany SLE. People suspected of having lupus are usually tested for anemia, leukopenia, and thrombocytopenia.

Anemia

Tests for anemia include those for hemoglobin, hematocrit, and red blood cell (RBC) count. In addition, the levels of iron, total iron-binding capacity, and ferritin may be tested. At any time during the course of the disease, about 40% of patients with SLE will be anemic. The anemia may be caused by iron deficiency, GI bleeding, medications, or autoantibody formation to RBCs. When first diagnosed, about 50% of patients have a form of anemia in which the concentration of hemoglobin and the size of the RBCs are normal. This is called normochromic-normocytic anemia, or "anemia of chronic disease." Autoimmune hemolytic anemia, with a positive Coombs test, is much less common.

Leukopenia and Thrombocytopenia

Abnormalities in the white blood cell (WBC) and platelet counts are an important indicator of SLE. Leukopenia, a decrease in the number of WBCs, is very common in active SLE and is found in 15–20% of patients. Thrombocytopenia, or a low platelet count, occurs in 25–35% of patients with SLE.

MEASUREMENTS OF AUTOIMMUNITY

The presence of certain autoantibodies have diagnostic value for SLE. The most specific tests are those that detect high levels of these autoantibodies. The most common and specific tests for autoantibodies and other elements of the immune system are listed first.

Antinuclear Antibody (ANA)

A screening test for ANA is standard in assessing SLE because it is positive in close to 100% of patients with active SLE. However, it is also positive in 95% of patients with mixed connective tissue disease, in more than 90% of patients with systemic sclerosis, in 70% of patients with primary Sjögren's syndrome, in 40–50% of patients with rheumatoid arthritis, and in 5–10% of patients with no systemic rheumatic disease. Patients with SLE tend to have high titers of ANA. False-positive results are found during chronic infectious diseases, such as subacute bacterial endocarditis,

tuberculosis, hepatitis, and malaria. The sensitivity and specificity of ANA determinations depend on the technique used.

Anti-Sm

Anti-Sm is an immunoglobulin specific against Sm, a ribonucleoprotein found in the cell nucleus. This test is highly specific for SLE; it is rarely found in patients with other rheumatic diseases. However, only 30% of patients with SLE have a positive anti-Sm test.

Anti-nDNA

Anti-nDNA is an immunoglobulin specific against native (double-stranded) DNA. This test is highly specific for SLE; it is not found in patients with other rheumatic diseases. Sixty to eighty percent of patients with active SLE have a positive anti-nDNA test. For many patients with anti-nDNA, the titer is a useful measure of disease activity. The presence of anti-nDNA is associated with a greater risk of lupus nephritis.

Anti-Ro(SSA) and Anti-La(SSB)

These immunoglobulins, commonly found together, are specific against RNA proteins. Anti-Ro is found in 30% of SLE patients and 70% of patients with primary Sjögren's syndrome. Anti-La is found in 15% of lupus patients and 60% of patients with primary Sjögren's syndrome. Anti-Ro is highly associated with photosensitivity; both are associated with neonatal lupus.

Complement

Complement proteins constitute a serum enzyme system that helps mediate inflammation. Complement components are triggered into an activated form by such immunologic events as interaction with immune complexes. Complement components are identified by numbers (C1, C2, etc.). Genetic deficiencies of C1q, C2, and C4, although rare, are commonly associated with SLE. A test to evaluate the entire complement system is called CH50. The most commonly measured complement components are the serum level of C3 and C4. These tests are particularly useful in evaluating kidney involvement and in monitoring the disease over time.

Erythrocyte Sedimentation Rate (ESR) and C-Reactive Protein (CRP)

Tests for ESR and CRP are nonspecific tests to detect generalized inflammation. Levels are generally increased in patients with active lupus and decline when corticosteroids or NSAIDs are used to reduce inflammation.

Antiphospholipid Antibodies (APLs)

APLs are autoantibodies that react with phospholipids. Recent data indicate that APLs recognize a number of phospholipid-binding plasma proteins (e.g., prothrombin, ß2-glycoprotein I) or protein-phospholipid complexes rather than phospholipids alone. APLs are present in 30–40% of lupus patients. A positive APL test plus the presence of arterial and venous thrombosis and thromboembolism or recurrent fetal deaths or thrombocytopenia is called APL syndrome. APL syndrome affects about a third of lupus patients with APLs (10–15% of all lupus patients). APLs and APL syndrome may also occur in patients without lupus (primary APL syndrome). APLs are detected in the following three types of laboratory assays.

Syphilis Serology

Certain blood tests for syphilis may be falsely positive in lupus patients. Chronically false-positive VDRL or rapid plasma reagin (RPR) tests may occur in patients with lupus. Cardiolipin, a phospholipid, is a component of the antigenic mixture used in these assays. More specific tests for syphilis, such as the fluorescent treponemal antibody-absorbed (FTA-ABS) and microhemagglutination-*Treponema pallidum* (MHA-TP) assays, are almost always negative in lupus patients without syphilis.

Anticardiolipin Antibody (ACA)

Sensitive enzyme-linked immunoabsorbent assays (ELISA) using cardiolipin as the putative antigen are commonly used to detect APLs. In patients with APL syndrome, most antibodies detected in anticardiolipin ELISAs are directed against cardiolipin-bound ß2-glycoprotein I.

Lupus Anticoagulant

Lupus anticoagulants are APLs that inhibit certain coagulation tests, such as the activated partial thromboplastin time (aPTT), dilute Russell viper venom time (dRVVT), and kaolin clotting time (KCT). Although the antibodies act as anticoagulants in these laboratory assays, they are not clinically associated with hemorrhage, but with thrombosis and other manifestations of the APL syndrome. Most lupus anticoagulant antibodies are directed against prothrombin or ß2-glycoprotein I.

TESTS FOR KIDNEY DISEASE

Several tests can be done to assess a patient for kidney disease.

Measurement of Glomerular Filtration Rate

The glomerular filtration rate is a measure of the efficiency of kidneys in filtering blood to excrete metabolic products. Typically this is done by collecting a 24-hour urine sample for measurement of creatinine clearance. Impairment of renal function by lupus nephritis results in reduced levels of creatinine clearances.

Urinalysis

Urinalysis can indicate the presence or extent of renal disease. For example, proteinuria can be a reliable indicator of renal disease. The presence of RBCs, WBCs, and cellular casts, particularly red cell casts, in the urine also indicates renal disease.

Measurement of Serum Creatinine Concentration

Creatinine is a waste product of muscle metabolism that is excreted by the kidney. Loss of renal function as a consequence of lupus nephritis causes increases in serum levels of creatinine. The concentration of creatinine in the serum can be used to assess the degree of renal impairment.

Kidney Biopsy

Kidney biopsy can be used to determine the presence of immune complexes and the presence, extent, and type of inflammation in the glomeruli. Diagnosis of the extent and type of inflammation may help to determine a treatment program for lupus.

Chapter Four

CARE OF THE LUPUS PATIENT

upus symptoms tend to present themselves according to the body system affected. These symptoms vary over time in intensity and duration for each patient as well as from patient to patient. To effectively care for a lupus patient, the nurse or other health professional needs an up-to-date knowledge and understanding of the disease, its many manifestations, and its changing and often unpredictable course.

This chapter provides an overview of general and system-specific lupus manifestations and identifies potential problems. Suggested health care interventions for the nonhospitalized lupus patient are given. Many of these interventions can be modified for the hospitalized patient. The information and nursing interventions described in this chapter are not meant to be inclusive, but to provide the practitioner with guidelines for developing a care plan specific to the needs of each lupus patient.

As a care plan is developed, the health professional should keep in mind the importance of frequently reassessing the patient's status over time and adjusting treatment to accommodate the variability of SLE manifestations. An additional and very important element of working with the lupus patient is to incorporate the patient's needs and routines in the plan of care. Adjusting nursing interventions and medical protocols to the patient's needs not only recognizes the value of the patient as an authority on her or his own illness but also can improve patient compliance and result in an improved quality of life.

Working together, the care provider and the patient have much to offer each other. The rewards are tremendous for the patient and family as independence is gained and the trust in the ability to care for oneself is strengthened.

SYSTEMIC LUPUS ERYTHEMATOSUS

General Manifestations

Fatigue, fever, psychological and emotional effects.

Specific Manifestations

Dermatologic: Butterfly rash, photosensitivity, DLE, subcutaneous LE, mucosal ulcers, alopecia, pain and discomfort, pruritus, bruising.

Musculoskeletal: Arthralgias, arthritis, other joint complications.

Hematologic: Anemia, decreased WBC count, thrombocytopenia, lupus anticoagulants, false-positive VDRL, elevated ESR.

Cardiopulmonary: Pericarditis, myocarditis, myocardial infarction, vasculitis, pleurisy, valvular heart disease.

Renal: Asymptomatic microscopic renal involvement, renal failure, fluid and electrolyte imbalance, urinary tract infection.

Central Nervous System (CNS): General CNS symptomology, cranial neuropathies, cognitive impairment, mental changes, seizures.

Gastrointestinal: Anorexia, ascites, pancreatitis, mesenteric or intestinal vasculitis.

Ophthalmologic: Eyelid problems, conjunctivitis, cytoid bodies, dry eyes, glaucoma, cataracts, retinal pigmentation.

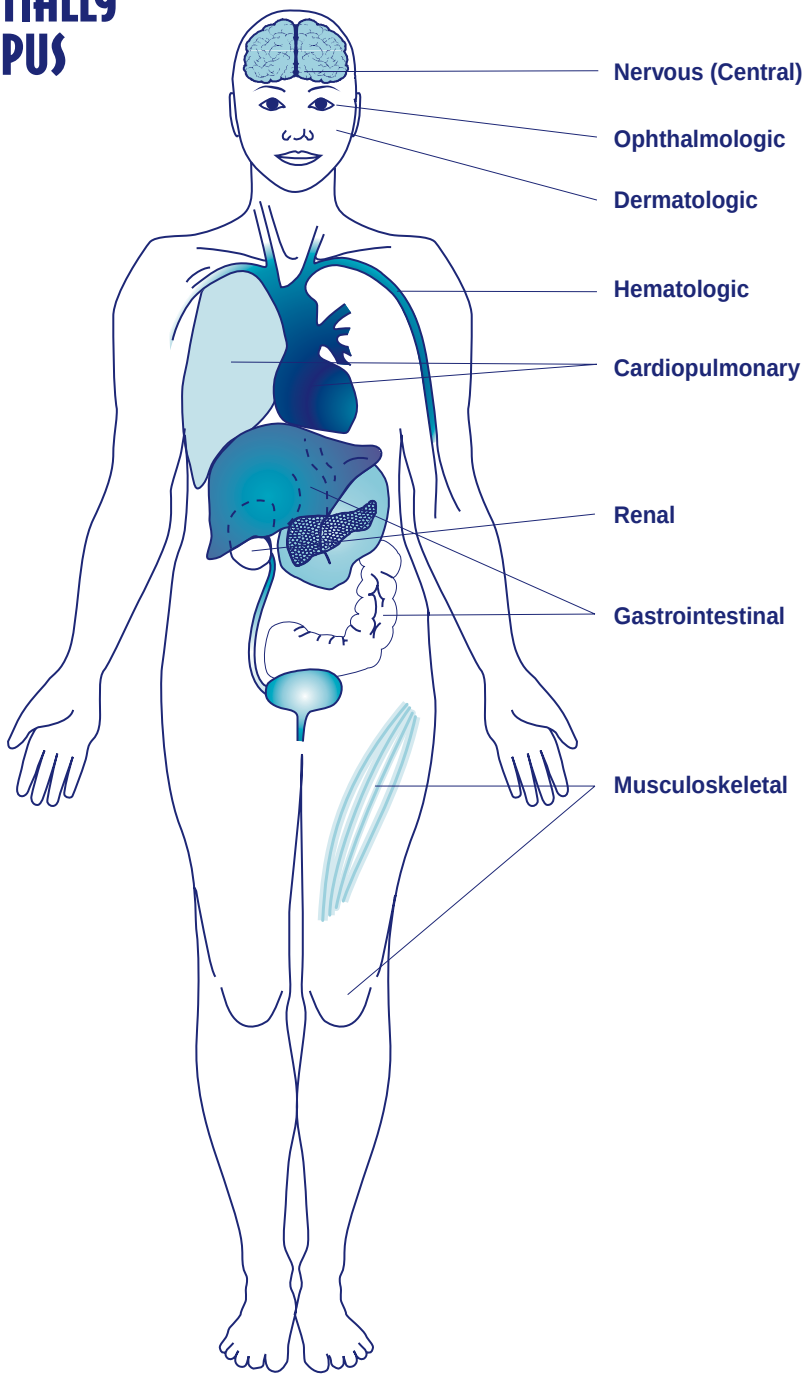
Other Key Issues

Pregnancy: Lupus flare, miscarriage or stillbirth, pregnancy-induced hypertension, neonatal lupus.

Infection: Increased risk of respiratory tract, urinary tract, and skin infections; opportunistic infections.

Nutrition: Weight changes; poor diet; appetite loss; problems with taking medications; increased risk of cardiovascular disease, diabetes, osteoporosis, and kidney disease.

SYSTEMS POTENTIALLY AFFECTED BY LUPUS



General Manifestations of SLE

Overview

Fatigue is a nearly universal complaint of patients with SLE even when no other manifestations of the disease are present. The cause of this debilitating fatigue is not known. The patient should be evaluated for factors that may exacerbate fatigue, such as overexertion, insomnia, depression, stress, anemia, and other inflammatory diseases. Fatigue in SLE patients may be lessened by adequate rest, healthful diet, exercise, and attention to psychosocial factors.

Many patients with SLE experience changes in weight. At least one-half of patients report weight loss before being diagnosed with SLE. Weight loss in SLE patients may be attributed to a decreased appetite, side effects of medications, gastrointestinal problems, or fever. Weight gain can occur in some patients and may be due in part to prescribed medications, especially corticosteroids, or fluid retention from kidney disease.

Episodic fever is experienced by more than 80% of SLE patients, and there is no particular fever pattern. Although high fevers can occur during a lupus flare, low-grade fevers are more frequently seen.

A complicating infection is often the cause of an elevated temperature in a patient with SLE. The patient's WBC count may be normal to elevated with an infection, but low with SLE alone. However, certain medications, such as immunosuppressives, will suppress the WBC even in the presence of fever. Therefore, it is important to rule out other causes of a fever, including an infection or a drug reaction. Urinary and respiratory infections are common in SLE patients.

Psychological and emotional effects, such as grief, depression, and anger, are commonly experienced by lupus patients. These can be related to the outward changes, such as skin alterations, caused by the disease as well as by other aspects of the disease and its treatment. It is important for health professionals to be alert to potential psychological repercussions and to assist in alleviating them.

Potential Physiological Manifestations

- Fatigue
- Weight gain or loss
- Fever — increased temperature over normal baseline
- Elevated WBC

POTENTIAL PSYCHOLOGICAL MANIFESTATIONS

- Lowered self-esteem
- Negative feelings about body
- Decreased confidence
- Feelings of decreased self-worth
- Depression
- Feelings of sadness, hopelessness, helplessness
- Difficulty in completing self-care activities, caring for children, maintaining a household, and other activities of daily living (ADL)
- Inability to maintain full- or part-time employment
- Decreased social activities
- Lack of energy or ambition
- Irritability
- Impaired concentration
- Crying
- Insomnia
- Suicidal thoughts

Potential Problems

1. Inability to complete activities of daily living (ADL) because of fatigue, weakness, and psychological difficulties
2. Changes in weight
3. Fever

Nursing Interventions

Objective: Minimize fatigue.

1. Assess patient's general fatigue level.
2. Assess for the presence of depression, anxiety, and other stressors.
3. Conduct assessment to determine patient's daily activities that contribute to fatigue.
4. Help patient to develop an energy-conserving plan for completing daily and other activities and work.
5. Suggest planning for rest periods as needed throughout the day to conserve energy.
6. Encourage patient to get 8–10 hours of sleep at night.
7. Encourage exercise as tolerated.

Objective: Maintain weight at optimal range.

1. Assess patient's prescription and non-prescription drug regimen and dosages.
2. Assess the patient's usual daily dietary intake by asking her or him to keep a food diary.
3. Develop a dietary plan with the patient that encourages healthful eating. If the patient has nutrition-related lupus complications, refer her or him to a registered dietitian for specialized counseling.
4. Encourage exercise as tolerated.
5. Record patient's weight at each visit.
6. Instruct patient to weigh herself or himself at home once a week and record it.

Objective: Teach patient to recognize fever and signs and symptoms of infection.

1. Assess patient's prescription and non-prescription drug regimen and dosages.
2. Monitor patient's WBC count.
3. Teach patient to monitor temperature during a lupus flare.

4. Teach patient to look for signs and symptoms of infection, particularly urinary and respiratory infections. (*Note:* The cardinal signs of infection may be masked because of corticosteroids and antipyretic medications.)
5. Instruct patient to call physician if signs and symptoms of an infection appear or if a fever is elevated above normal baseline.

Objective: Assist patient in adjusting to physical and lifestyle changes.

1. Allow patient to express feelings and needs.
2. Assess patient's usual coping mechanisms.
3. Acknowledge that feelings of denial and anger are normal.
4. Explore with patient sources of potential support and community resources.
5. Explore possible ways of concealing skin lesions and hair loss.
6. Encourage patient to discuss interpersonal and social conflicts that arise.
7. Encourage patient to accept help from others, such as counseling or a support group.

Objective: Recognize the signs and symptoms of depression and initiate a plan of care.

1. Assess patient for the major signs and symptoms of depression.
2. Assess patient's interpersonal and social support systems.
3. Encourage patient to express feelings.
4. Initiate a referral to a mental health counselor or psychiatrist.

Note: For additional information, see the Patient Information Sheets (Chapter 7) on Living With Lupus and Skin Care and Lupus.

For further information and nursing interventions, see the section on infection in this chapter, pages 52–53. Also see the Patient Information Sheets (Chapter 7) on Living With Lupus, Preventing Fatigue Due to Lupus, and Fever and Lupus.

DERMATOLOGIC MANIFESTATIONS

Overview

Approximately 80% of patients with SLE have skin manifestations and often suffer from itching, pain, and disfigurement. The classic sign of SLE is the "butterfly" rash extending over the cheeks (malar area) and bridge of the nose. This rash ranges from a faint blush to a severe eruption with scaling. It is photosensitive, and it may be transitory or fixed. Between 55 and 85% of patients develop this rash at some time in the course of the disease.

Other rashes may occur elsewhere on the face and ears, upper arms, shoulders, chest, and hands. DLE is seen in 15–30% of patients with SLE. Subacute cutaneous LE, seen in about 10% of SLE patients, produces highly photosensitive papules that itch and burn. Skin changes, especially the butterfly rash and subacute cutaneous LE, can be precipitated by sunlight.

Some patients may develop mouth, vaginal, or nasal ulcers. Hair loss (alopecia) occurs in about one-half of SLE patients. Most hair loss is diffuse, but it may be patchy. It can be scarring or nonscarring. Alopecia may also be caused by corticosteroids, infection, or immunosuppressive drugs.

Raynaud's phenomenon (paroxysmal vasospasm of the fingers and toes) frequently occurs in patients with SLE. For most patients, Raynaud's phenomenon is mild. However, some SLE patients with severe Raynaud's phenomenon may develop painful skin ulcers or gangrene on the fingers or toes.

Varying levels of pain and discomfort due to skin alterations may occur. Pruritus accompanies many types of skin lesions. Attacks of Raynaud's phenomenon can cause a deep tingling feeling in the hands and feet that can be very uncomfortable. Both pain and itching may affect a patient's ability to carry out activities of daily living (ADL).

Skin alterations in the lupus patient, particularly those of DLE, can be disfiguring. As a result, patients may experience fear of rejection by others, negative feelings about their body, and depression. Changes in lifestyle and social involvement may occur.

Potential Problems

1. Alteration in skin integrity
2. Alopecia
3. Discomfort (pain, itching)
4. Alteration in body image
5. Depression

POTENTIAL DERMATOLOGIC MANIFESTATIONS

- ▲ Butterfly rash on cheeks and bridge of nose
- ▲ Scaly, disk-shaped scarring rash (DLE)
- ▲ Erythematous, slightly scaly papules (subacute cutaneous LE)
- ▲ Psoriasiform or arcuate (curved) lesions on the trunk of the body (subacute cutaneous LE)
- ▲ Itching and burning
- ▲ Ulcers in the mouth, vagina, or nasal septum
- ▲ Atrophy (including striae or stretch marks)
- ▲ Impaired wound healing
- ▲ Easy bruising
- ▲ Petechiae
- ▲ Increased body hair (hirsutism)
- ▲ Steroid-induced ecchymosis
- ▲ Ulcers or gangrene on fingers or toes
- ▲ Alopecia

Nursing Interventions

Objective: Minimize appearance of lesions.

1. Document appearance and duration of lesions and rashes.
2. Teach patient to minimize direct exposure to UV rays from sun and from fluorescent and halogen light bulbs. (Glass does not provide complete protection from UV rays.)
3. Instruct patient to use a sunscreen with an SPF of 15 or greater and wear protective clothing. Patients who are allergic to PABA will need to find a PABA-free sunscreen.
4. Provide information on hypoallergenic concealing makeup.
5. Instruct patient to avoid topical applications, such as hair dyes and skin creams, and the use of certain drugs that may make her or him more sensitive to the sun.

Objective: Alleviate discomfort.

1. For patients with mouth lesions, suggest a soft-food diet, lip balms, and warm saline rinses.
2. Instruct patient to take medications that may help to alleviate discomfort and itching as ordered. (The doctor may give the patient intralesional steroid injections.)
3. Suggest self-help measures for patients with Raynaud's phenomenon, including:
   - keep warm, particularly in cold weather; use chemical warmers, gloves, socks, hats; avoid air conditioning; use insulated drinking glasses for cold drinks; wear gloves when handling frozen or refrigerated foods;
   - quit smoking;
   - control stress; and
   - exercise as tolerated.

Objective: Help patients to cope with potential psychological manifestations.

1. See the nursing interventions dealing with psychological issues under manifestions on pages 31–32.

Overview

Arthralgia or arthritis is experienced by 95% of SLE patients at some time during the course of the disease. Articular pain is the initial symptom in about one-half of patients eventually diagnosed with SLE. Morning stiffness and joint and muscle aching can also occur. Joint pain may be migratory; it is typically symmetric but is asymmetric in many patients. Joints may become warm and swollen. X rays of the joints usually do not show erosion or destruction of bone.

Unlike rheumatoid arthritis, the arthritis of SLE tends to be transitory. Proliferation of the synovium is more limited, and joint destruction is rare. The joints most commonly involved are those of the fingers, wrists, and knees; less commonly involved are the elbows, ankles, and shoulders.

Several joint complications may occur in SLE patients, including Jaccoud's arthropathy and osteonecrosis. Subcutaneous nodules, especially in the small joints of the hands, are seen in about 5% of patients. Tendinitis, tendon rupture, and carpal tunnel syndrome are seen occasionally.

Potential Problems

1. Pain
2. Alteration in joint function

Nursing Interventions

Objective: Minimize pain from joint and muscle complications.

1. Assess and document joint complaints and appearance. Changes may be transient.
2. Assess patient's self-management techniques for controlling pain.
3. Teach patient to apply heat or cold as appropriate.
4. Instruct patient in use of prescription and nonprescription pain medications.
5. If ordered by physician, teach patient to apply splints or braces.

POTENTIAL MUSCULOSKELETAL MANIFESTATIONS

- Morning stiffness and aching
- Joint pain
- Warm, swollen joints
- Ulnar deviation of the fingers with swan neck deformities and subluxations
- Generalized myalgia and muscle tenderness, especially in the upper arms and upper legs

Objective: Maintain joint function and increase muscle strength.

1. Suggest warm showers or baths to lessen stiffness and pain.
2. If indicated, refer patients with acutely inflamed joints to a physical therapist for passive range-of-motion (ROM) exercises. The physical therapist may train a family member to assist the patient with ROM exercises at home.
3. Teach patient that an inflamed joint should not bear weight and suggest that patient avoid strenuous activity.
4. If needed, assist patient to obtain crutches, a walker, or a cane.
5. Assist patient in developing a regular exercise plan that can be carried out during periods of remission. This plan should include exercises that promote muscle tone and fitness, minimize fatigue, and increase well-being.
6. Consider referring patient to an occupational therapist.

Note: For additional information, see the Patient Information Sheets (Chapter 7) on Exercise and Lupus, Preventing a Lupus Flare, and Joint Function and Lupus.

HEMATOLOGIC MANIFESTATIONS

Overview

Abnormal blood conditions are common in patients with SLE. Problems include anemia, thrombocytopenia, and other clotting disorders.

Anemia, which is common in SLE patients, reflects insufficient bone marrow activity, shortened RBC life span, or poor iron uptake. Aspirin, NSAIDs, and prednisone can cause stomach bleeding and exacerbate the condition. There is no specific therapy for this type of anemia. Immune-mediated anemia (or hemolytic anemia), which is due to antibodies directed at RBCs, is treated with corticosteroids.

Thrombocytopenia may occur and may respond to low-dose corticosteroids. Mild forms may not need to be treated, but a severe form requires high-dose corticosteroid or cytotoxic drugs. The major clinical features of APLs and APL syndrome are venous thrombosis, arterial thrombosis, and thrombocytopenia with a history of positive anticardiolipin antibody (ACL) tests.

Abnormal laboratory tests may include a false-positive VDRL test for syphilis. Fluorescent treponemal antibody absorption (FTA-ABS) and microhemagglutination-*Treponema pallidum* (MHA-TP) tests, which are more specific tests for syphilis, are almost always negative if the patient does not have syphilis. An elevated erythrocyte sedimentation rate (ESR) is a common finding in active SLE, but it does not always mirror disease activity.

Potential Problems

1. Inability to complete ADL because of fatigue and weakness
2. Anemia
3. Potential for hemorrhage
4. Potential to develop venous or arterial thromboses
5. Increased risk of infection

Nursing Interventions

Objective: Minimize fatigue.

1. Refer to the nursing interventions for fatigue on page 31.

Objective: Recognize anemia and develop plan of care.

1. Monitor patient for signs and symptoms of anemia and for altered laboratory values.

POTENTIAL HEMATOLOGIC MANIFESTATIONS

Anemia

- ▲ Decreased hemoglobin and hematocrit values
- ▲ Positive Coombs' test (hemolytic anemia)
- ▲ Tachycardia
- ▲ Palpitations
- ▲ Dizziness
- ▲ Sensitivity to cold
- ▲ Chronic fatigue, lethargy, and malaise
- ▲ Pallor
- ▲ Weakness
- ▲ Dyspnea on exertion
- ▲ Headache

Thrombocytopenia

- ▲ Petechiae
- ▲ Excessive bruising of skin
- ▲ Bleeding from gums, nose
- ▲ Blood in stool

2. Develop a plan with patient to conserve energy.
3. Teach patient the basics of good nutrition.
4. Instruct patient to take iron preparation medications as prescribed.

Objective: Minimize episodes of bleeding.

1. Assess patient for signs and symptoms of bleeding, such as petechiae, bruises, GI bleeding, blood in urine, ecchymoses, nose bleeds, bleeding from the gums, heavy menses, and bleeding between menstrual periods.
2. Teach patient why she or he is at risk of bleeding (low platelet count, anemia, thrombocytopenia) and to report episodes to physician.
3. Encourage patient to wear a medical alert bracelet or carry a card.
4. Teach patient measures to prevent bleeding, such as use of a soft toothbrush or an electric shaver.

Objective: Decrease risk of infection.

1. See the nursing interventions for infection on pages 52–53.

Note: For more information, see Laboratory Tests Used to Diagnose and Evaluate SLE (Chapter 3) and the Patient Information Sheet (Chapter 7) on Preventing Fatigue Due to Lupus.

CARDIOPULMONARY MANIFESTATIONS

Overview

Cardiac abnormalities contribute significantly to morbidity and mortality in SLE and are one of the most important clinical manifestations of the disease. In addition, involvement of the lungs and pleurae is common. Pericarditis, an inflammation of the pericardium, is the most common cardiac abnormality in SLE. Myocarditis, an inflammation of the heart muscle, may also occur, but is rare. Myocardial infarction, caused by atherosclerosis, has been reported in SLE patients below the age of 35 years.

Vasculitis (inflammation of the blood vessels) and serositis (inflammation of serous membranes) are frequently part of the autoimmune pathology of SLE. These conditions respond well to corticosteroids. Vasculitis may cause many different symptoms, depending on the system(s) most affected. Serositis most commonly presents as pleurisy or pericarditis. Pleuritic chest pain is common. Pleurisy is the most common respiratory manifestation in SLE. Attacks of pleuritic pain can also be associated with pleural effusions. Many patients complain of chest pain, but pericardial changes are not often demonstrated on clinical evaluation.

Potential Problems

1. Alterations in cardiac function
2. Potential for impaired gas exchange and ineffective breathing patterns
3. Alteration in tissue perfusion

POTENTIAL CARDIOPULMONARY MANIFESTATIONS

Pericarditis

- ▲ Pain in the anterior chest, neck, back, or arms that is often relieved by sitting up
- ▲ Shortness of breath
- ▲ Swelling of legs and feet
- ▲ Fever
- ▲ Chills
- ▲ Audible pericardial friction rub

Myocarditis

- ▲ Chest pain
- ▲ Shortness of breath
- ▲ Fever
- ▲ Fatigue
- ▲ Palpitations

Atherosclerosis Leading to Myocardial Infarction

Warning signs of myocardial infarction:

- ▲ Burning, choking, squeezing, or pressing chest pain that may radiate to left shoulder and arm
- ▲ Shortness of breath
- ▲ Weakness
- ▲ Unrelieved indigestion
- ▲ Nausea and vomiting

Pleurisy

- ▲ Shortness of breath
- ▲ Chest pain, especially with deep inspiration
- ▲ Coughing up blood or thick mucus

Continued on next page

POTENTIAL CARDIOPULMONARY MANIFESTATIONS (CONTINUED)

Periungual Erythema
- ▲ Redness in the nailbed

Livedo Reticularis
- ▲ A reddish or cyanotic pattern seen on arms, legs, torso, especially in cold weather

Leukocytoclastic Vasculitis
- ▲ Necrotic ulcerations, including raised hemorrhagic nodules (papule, purpura) that ulcerate, especially on the lower legs, ankles, and dorsa of the feet

Valvular Heart Disease (Libman-Sacks Lesions)
- ▲ Lesions that may result in cardiac murmurs and valve dysfunction; associated with antiphospholipid antibodies

Venous Thrombosis
- ▲ Positive Homans' sign
- ▲ Pain, swelling, inflammation, redness, and warmth in the affected limb
- ▲ Increased circumference of affected limb

Arterial Thrombosis
- ▲ Pain or loss of sensation due to ischemia
- ▲ Panesthesias and loss of position sense
- ▲ Coldness
- ▲ Pallor
- ▲ Paralysis
- ▲ No pulse

Nursing Interventions

Objective: Detect changes in cardiac function.

1. Assess patient for signs and symptoms of potential cardiac problems.
2. Teach patient signs and symptoms of cardiac problems, including warning signs of a heart attack; reinforce the importance of reporting them to the physician.
3. Educate patient about medications.
4. Educate patient about a healthful diet and regular exercise as tolerated.

Objective: Maintain adequate gas exchange and effective breathing patterns.

1. Assess quality and depth of respirations; auscultate breath sounds.
2. Suggest measures to relieve pain, such as relaxation techniques, biofeedback, rest, and pain medications as ordered.
3. Encourage patients who smoke to quit.

Objective: Ensure adequate tissue perfusion.

1. Assess skin color and temperature; check for lesions.
2. Check capillary refill in the nailbeds.
3. Assess for presence of edema and pain in the extremities.
4. Stress the importance of not smoking.
5. Teach patient the basics of good foot care.
6. Teach patient to avoid cold temperatures and to keep the hands and feet warm, especially in winter months.
7. Teach patient signs and symptoms of vascular impairment that need to be reported to the physician, including a change in skin color or sensation or appearance of lesions.

Objective: Recognize the signs and symptoms of thromboses; refer for immediate medical attention.

1. Teach patient the signs and symptoms of potential venous or arterial thrombosis and reinforce the need to contact a physician immediately.

Note: For additional information, see the Patient Information Sheet (Chapter 7) on Serious Conditions Associated With Lupus.

RENAL MANIFESTATIONS

Overview

Renal damage is one of the most serious complications of SLE. The majority of lupus patients have some degree of asymptomatic microscopic kidney damage. Less than 50% have clinical renal disease, and most of those with renal disease have one of the milder forms. Kidney damage may necessitate treatment with corticosteroids, cytotoxic agents, dialysis, or renal transplantation.

Renal biopsy can be helpful in making decisions about drug treatments and determining prognosis by assessing the presence of active renal disease versus scarring.

Potential Problems

1. Impaired renal function
2. Fluid and electrolyte imbalance
3. Increased risk of infection

POTENTIAL RENAL MANIFESTATIONS

SLE Nephropathy

Signs and symptoms:

- Hematuria (as few as 5 RBCs is significant)
- Proteinuria (>1+ to 2+)
- Abacterial pyuria
- Elevated creatinine level (indicates loss of renal function)
- Elevated blood urea nitrogen (BUN)
- Markedly abnormal serologic tests, such as decreased complement or elevated anti-DNA values
- Weight gain
- Ankle edema
- Hypertension

Signs and symptoms suggesting renal failure

- Nausea and vomiting
- Anorexia
- Anemia
- Lethargy
- Pruritus
- Changing level of consciousness

Fluid and Electrolyte Imbalance (Excess Extracellular Fluid Volume)

- Weight gain
- Pitting edema of the lower extremities
- Sacral edema
- Bounding pulse, elevated blood pressure, S3 gallop
- Engorgement of neck and hand veins
- Dyspnea
- Constant cough
- Crackles in lungs
- Cyanosis
- Decreased hematocrit
- Urine specific gravity <1.010
- Variable serum sodium level (normal, high, or low), depending on the amount of sodium retention or water retention
- Serum osmolality <275 mOsm/kg

Continued on next page

POTENTIAL RENAL MANIFESTATIONS (CONTINUED)

Urinary Tract Infection

- ▲ Dysuria
- ▲ Frequent urination
- ▲ Urgent need to urinate
- ▲ Fever
- ▲ Cloudy urine
- ▲ Incomplete emptying of the bladder
- ▲ Low back or suprapubic pain
- ▲ Flank pain
- ▲ Malaise
- ▲ Nausea and vomiting

Nursing Interventions

Objective: Promptly recognize renal involvement and prevent complications.

1. Document any patient complaints or assessment findings that may indicate renal involvement.
2. Teach patient to watch for signs and symptoms of renal complications and report them promptly to the physician: headache, facial swelling, peripheral edema, dizziness, "foamy" urine (proteinuria), "coke-colored" urine (hematuria), or nocturia and urinary frequency.
3. Assess patient for early signs of heart or liver failure.
4. Refer patient to a dietitian for counseling on dietary changes to accommodate alterations in renal status.
5. Teach patient to take prescribed medications as ordered.
6. Stress the importance of referral and followup care with nephrologist if necessary.

Objective: Decrease fluid retention and edema.

1. Monitor electrolyte values.
2. Assess breath sounds and instruct patient to report shortness of breath or dyspnea.
3. Teach patient to maintain balanced fluid intake and output.
4. Monitor patient for signs and symptoms of extracellular fluid overload.
5. Instruct patient to weigh herself or himself daily to monitor fluid retention.
6. Monitor patient's blood pressure and teach patient how to monitor it at home.

Objective: Minimize risk of infection.

1. Teach patient to watch for the signs and symptoms of urinary tract infection and to report them to the physician.
2. Instruct patient that corticosteroid therapy may mask the usual symptoms of infection and that she or he may have an altered immune response because of medications used to control SLE.
3. Teach patient to take antibiotics for urinary tract infection as prescribed.

Note: For additional information, see the Patient Information Sheet (Chapter 7) on Serious Conditions Associated With Lupus.

CENTRAL NERVOUS SYSTEM MANIFESTATIONS

Overview

Neurologic manifestations of SLE are common and vary from mild to severe. They can be difficult to diagnose and distinguish from other diseases. All portions of the nervous system may be affected, including the CNS. Definite diagnosis of CNS lupus may be difficult, as symptoms may be related to medications, other medical conditions, or to individual reactions to chronic illness.

Cranial or peripheral neuropathy occurs in 10–15% of patients; it is probably secondary to vasculitis in small arteries supplying nerves. Cerebrovascular accidents (strokes) are reported in approximately 15% of patients. Between 10 and 20% of patients experience seizures. Although cognitive impairment is believed to be very common, there are few measurements to document it.

Serious CNS involvement ranks behind only kidney disease and infection as a leading cause of death in lupus. However, the majority of SLE patients with CNS complications do not develop a life-threatening disease.

Potential Problems

1. Alteration in mental status, cognition, and perception
2. Altered ability to perform ADL and meet family responsibilities
3. Potential for injury

POTENTIAL CNS MANIFESTATIONS

General CNS lupus

- ▲ Headaches
- ▲ Fever
- ▲ Confusion
- ▲ Seizures
- ▲ Psychosis

Cranial neuropathies

- ▲ Visual defects
- ▲ Blindness
- ▲ Nystagmus (involuntary movement of the eyeball)
- ▲ Ptosis (paralytic drooping of the eyelid)
- ▲ Papilledema (edema in the optic disk)
- ▲ Tinnitus
- ▲ Vertigo
- ▲ Facial palsy

Cognitive impairment

- ▲ Confusion
- ▲ Impaired long- and short-term memory
- ▲ Difficulty in conceptualizing, abstracting, generalizing, organizing, and planning information for problem solving
- ▲ Difficulties in personal and extrapersonal orientation

Continued on next page

POTENTIAL CNS MANIFESTATIONS (CONTINUED)

- ▲ Altered visual-spatial abilities
- ▲ Selective attention
- ▲ Difficulties in pattern recognition, sound discrimination and analysis, and visual-motor integration

Mental changes

- ▲ Depression
- ▲ Anxiety
- ▲ Affective disorder
- ▲ Mood swings
- ▲ Hypomania or mania (especially with corticosteroid use)

Rare CNS manifestations

- ▲ Movement disorder
- ▲ Aphasia
- ▲ Coma

Nursing Interventions

Objective: Develop plan for patient to perform ADL appropriately and independently.

1. Assess and document patient's mental status to determine her or his capabilities:
   - general appearance;
   - unusual body movements;
   - speech patterns and word use;
   - alertness and orientation to time, place, and person;
   - memory of remote and recent past;
   - perception of self and environment;
   - affect and emotional stability;
   - ability to solve problems; and
   - presence of depression.
2. Support patient's need to maintain some control over daily activities and decisions:
   - encourage patient to plan and participate in daily routines;
   - set aside time to develop trust and rapport with patient, and be consistently truthful (patients are keenly aware of inconsistencies in information provided).
3. Encourage patient to discuss effects of SLE on her or his personal life and coping methods. Allow expressions of fear and anger.

Objective: Assist patient in identifying family and community support services.

1. Assess patient's support network. Discuss alternatives for strengthening supports.
2. Anticipate family concerns. Seek out the family to answer their questions and to provide support. Include significant others in patient care as appropriate.
3. Help family identify potential coping skills, environmental supports, and community services for dealing with chronically ill people.
4. Encourage patient and family members to consider professional counseling.

Objective: Minimize potential for injury.

1. Assist patient and family in identifying and removing potentially dangerous items in the environment.
2. Involve family members in planning of patient's care and safety measures.
3. Assess patient's ability to safely administer own medications.

Note: For additional information, see the Patient Information Sheets (Chapter 7) on Living With Lupus and Serious Conditions Associated With Lupus.

Overview

Gastrointestinal (GI) problems are common and range from vague complaints of anorexia to life-threatening bowel perforation secondary to mesenteric arteritis. Anorexia, nausea, vomiting, and diarrhea may be related to the use of salicylates, NSAIDs, antimalarials, corticosteroids, and cytotoxic drugs.

SLE patients who present with acute abdominal pain and tenderness need immediate, aggressive, and comprehensive evaluation to rule out an intra-abdominal crisis. Ascites, an abnormal accumulation of fluid in the peritoneal cavity, is found in about 10% of SLE patients. Pancreatitis is a serious complication occurring in approximately 5% of SLE patients and is usually secondary to vasculitis.

Mesenteric or intestinal vasculitis is a life-threatening condition that may have complications of obstruction, perforation, or infarction. They are seen in more than 5% of patients with SLE. Abnormal liver enzyme levels are also found in about one-half of SLE patients (usually secondary to medications). Active liver disease is rarely found.

POTENTIAL GI MANIFESTATIONS

General manifestations
- Persistent sore throat
- Dry mouth (characteristic of patients with coexisting Sjögren's syndrome)
- Anorexia
- Nausea and vomiting
- Diarrhea
- Dysphagia (especially in association with Raynaud's phenomenon)

Pancreatitis
- Mild nonspecific abdominal pain to severe epigastric pain radiating to the back
- Nausea
- Vomiting
- Elevated serum amylase level
- Dehydration

Ascites
- Abdominal distention
- Bulging flanks
- Downward protruding umbilicus

Mesenteric and intestinal vasculitis
- Cramping or constant abdominal pain
- Vomiting
- Fever
- Diffuse direct and rebound abdominal tenderness

Nutritional Deficiencies
See pages 54–55 for signs and symptoms of nutritional deficiencies.

Potential Problems

1. Alteration in GI function related to drug therapy or disease process
2. Nutritional deficiencies

Nursing Interventions

Objective: Minimize GI side effects from medications.

1. See Chapter 5, *Medications Used to Treat Lupus* and the Patient Information Sheets (Chapter 7) on Nonsteroidal Anti-Inflammatory Drugs, Antimalarials, Corticosteroids, Azathioprine, and Cyclophosphamide.

Objective: Minimize complications from GI manifestations.

1. Assess patient for GI problems at each visit.
2. Monitor laboratory results.
3. Suggest measures that may increase comfort, such as throat lozenges, saline rinses, or small, frequent meals.
4. Instruct patient to immediately report any sudden or severe abdominal pain, shortness of breath, or epigastric pain to physician.
5. Refer patient to dietitian.

Objective: Maintain nutritional status.

1. See the nursing interventions under the nutrition section of this chapter (pages 54–55) and the Patient Information Sheet on Nutrition and Lupus.

OPHTHALMOLOGIC MANIFESTATIONS

Overview

Visual impairment may be due to SLE or to drug treatment (corticosteroids or antimalarials), or it may be a separate problem (glaucoma or retinal detachment). Blindness due to SLE occurs, but is rare. Other visual problems may occur:

- A lupus rash may develop on the eyelids.
- Conjunctivitis occurs in 10% of SLE patients and is usually infectious. Kerato-conjunctivitis is usually mild.
- Cytoid bodies are the most common retinal change in SLE. They reflect microangiopathy of the retinal capillaries and localized microinfarction of the superficial nerve fiber layers of the retina.
- Sjögren's syndrome is an autoimmune condition manifest as excessive dryness of mucous membranes. Lupus patients with these symptoms require artifical tears to relieve dry eyes.
- Glaucoma and cataracts may be caused by corticosteroids.
- Antimalarials can damage the retina, which can impair vision (particularly color vision) or, rarely, cause blindness.

Potential Problems

1. Discomfort
2. Visual impairment
3. Potential for injury
4. Difficulty carrying out ADL

Nursing Interventions

Objective: Minimize discomfort.

1. Allow time for patient to express concerns and ask questions.
2. Teach the patient how to apply artificial tears for dry eyes to increase comfort and prevent corneal abrasion.
3. Teach patient the correct way to take prescribed medications, such as eye drops for glaucoma.
4. Suggest warm, moist compresses, which may help ease discomfort and itching from conjunctivitis.

Objective: Minimize potential for serious visual impairment or blindness.

1. Assess patient's vision changes and impairments.
2. Reinforce the need to follow up with an ophthalmologist.

POTENTIAL OPHTHALMOLOGIC MANIFESTATIONS

- A lupus rash on the eyelids
- Red, sore, swollen eyes
- Tearing
- Mucus discharge from eyes, particularly upon awakening
- Sensitivity to light
- Change in vision
- Blurred vision
- Cloudy lens(es)
- Dry eyes
- Burning sensation in eyes

Objective: Develop a plan for patient to perform ADL appropriately and independently.

1. Provide referrals to support groups and services for the visually impaired.
2. Refer to the CNS lupus nursing interventions on page 45 for additional suggestions.

Objective: Minimize potential for injury.

1. See the CNS lupus nursing interventions on page 45 for suggestions.

Note: For additional information, see the Patient Information Sheets (Chapter 7) on Living With Lupus and Serious Conditions Associated With Lupus.

PREGNANCY

Overview

Twenty years ago, women with lupus were counseled not to become pregnant because of the risk of a flare of the disease and an increased risk of miscarriage. Research and careful treatment have made it possible for more and more women with lupus to have successful pregnancies. Although a lupus pregnancy is still considered high risk, most women with lupus are able to carry their babies safely to term. Experts disagree on the exact numbers, but approximately 20–25% of lupus pregnancies end in miscarriage, compared with 10–15% of pregnancies in women without the disease. Pregnancy counseling and planning before pregnancy is important. Optimally, a woman should have no signs or symptoms of lupus before she becomes pregnant.

Researchers have now identified two closely related lupus autoantibodies, anticardiolipin antibody and lupus anticoagulant, that are associated with risk of miscarriage. One-third to one-half of women with lupus have these autoantibodies, which can be detected by blood tests. Identifying women with the autoantibodies early in the pregnancy may help physicians take steps to reduce the risk of miscarriage. Pregnant women who test positive for these autoantibodies and who have had previous miscarriages are generally treated with baby aspirin or heparin throughout their pregnancy.

Some women may experience a mild to moderate flare during or after their pregnancy; others may not. Pregnant women with lupus, especially those taking corticosteroids, are also likely to develop pregnancy-induced hypertension, diabetes, hyperglycemia, and kidney complications. About 25% of babies of women with lupus are born prematurely, but do not suffer from birth defects.

About 3% of babies born to mothers with SLE will have neonatal lupus, or specific antibodies called anti-Ro(SSA) and anti-La(SSB). This is not the same as SLE and is almost always temporary. The syndrome is thought to be caused by passive transfer of anti-Ro antibodies from the mother to the fetus. About one-third of women with SLE have this antibody. By 3–6 months of age, the rash and blood abnormalities associated with neonatal lupus disappear. Very rarely, babies with neonatal lupus will have a congenital complete heart block. This problem is permanent, but can be treated with a pacemaker.

POTENTIAL LUPUS COMPLICATIONS DURING PREGNANCY

Lupus flare
- ▲ Increased pain
- ▲ Morning stiffness
- ▲ Fever
- ▲ Development or worsening of a rash
- ▲ Stomach discomfort
- ▲ Headache
- ▲ Dizziness

Miscarriage
- ▲ Cramping
- ▲ Vaginal bleeding (spotting to heavy bleeding)

Pregnancy-induced hypertension

Mild
- ▲ Blood pressure 140/90 and over during the second half of pregnancy
- ▲ Mild, generalized edema
- ▲ Proteinuria

Pre-eclampsia
- ▲ Blood pressure 140/90 and over during the second half of pregnancy
- ▲ Proteinuria
- ▲ Epigastric pain
- ▲ Hyperreflexia
- ▲ Edema, including face and hands
- ▲ Headache

Eclampsia
- ▲ All of the symptoms of preeclampsia
- ▲ Seizures

Neonatal lupus
- ▲ Transient rash
- ▲ Transient blood count abnormalities
- ▲ Heartblock

Potential Problems

1. Lupus flare
2. Increased risk of spontaneous abortion or stillbirth
3. Pregnancy-induced hypertension
4. Increased risk of prematurity
5. Neonatal lupus

Nursing Interventions

Objective: Educate the woman regarding birth control options and risks of pregnancy.

1. Encourage patient to plan pregnancy during remission and only after consulting with her doctor.
2. Discuss birth control options:
   - Barrier methods (diaphragm or condom with spermicidal foam) are the safest.
   - IUDs are not recommended because of increased risk of infection.
   - Oral contraceptives may be appropriate.
3. Discuss the potential risks of pregnancy and the importance of careful monitoring.

Objective: Ensure a healthy, full-term pregnancy.

1. Urge patient to keep appointments with her primary doctor and obstetrician.
2. Instruct patient to observe for signs of complications or an impending flare.
3. Monitor blood pressure and watch for signs of toxemia, which may be hard to distinguish from a lupus flare.

Note: For additional information, see the Patient Information Sheet (Chapter 7) on Pregnancy and Lupus.

INFECTION

Overview

SLE affects the immune system, thus reducing the body's ability to prevent and fight infection. In addition, many of the drugs used to treat SLE also suppress the function of the immune system, thereby further depressing the ability to fight infection. The risk of infection parallels medication dosages and duration of treatment.

Patients with SLE who show signs and symptoms of infection need prompt therapy to prevent it from becoming life threatening. The most common infections involve the respiratory tract, urinary tract, and skin and do not require hospitalization if they are treated promptly. Other opportunistic infections, particularly *Salmonella*, herpes zoster, and *Candida* infections, are more common in patients with SLE because of altered immune status.

Potential Problems

1. Increased risk of infection

POTENTIAL MANIFESTATIONS OF INFECTION

Respiratory tract infections

- ▲ Sore throat
- ▲ Sneezing
- ▲ Fever
- ▲ Productive or nonproductive cough
- ▲ Runny nose
- ▲ Malaise
- ▲ Chills
- ▲ Back and muscle pain
- ▲ Dyspnea
- ▲ Wheezing or rales
- ▲ Chills
- ▲ Nausea
- ▲ Vomiting

Urinary tract infections

- ▲ Chills
- ▲ Fever
- ▲ Flank pain
- ▲ Nausea
- ▲ Vomiting
- ▲ Urinary frequency
- ▲ Dysuria
- ▲ Hematuria

Skin infections

- ▲ Lesions
- ▲ Redness
- ▲ Swelling
- ▲ Tenderness or pain

Nursing Interventions

Objective: Minimize incidence of infection.

1. Assess patient's current medications, particularly those that promote susceptibility to infection such as corticosteroids and immunosuppressives.
2. Teach patient to use good hand-washing and personal-hygiene techniques.
3. Teach patient the signs and symptoms of infection and reinforce the importance of reporting them to the physician.
4. Encourage patient to eat a balanced diet with adequate calories to help preserve the immune system.
5. Teach patient to minimize exposure to crowds and people with infections or contagious illnesses.

Objective: Educate the patient about immunizations.

1. Check patient's current immunization status.
2. Teach patient that infections can be minimized with immunizations.
3. Encourage patient to consult her or his doctor before considering allergy shots or flu or pneumococcal vaccines; these medications may induce a lupus flare.

Note: For additional information, see the section on general manifestations of SLE in this chapter, pages 30–31. Also see the Patient Information Sheet (Chapter 7) on Fever and Lupus.

NUTRITION

Overview

The patient with lupus often has special nutritional needs related to medical conditions that may arise during the course of the disease. These conditions include steroid-induced osteoporosis or diabetes, cardiovascular disease, and kidney disease. For the SLE patient to maintain optimal health, the nurse must work closely with the patient, dietitian, and physician to develop a nutritional plan specific to the patient's disease and manifestations.

POTENTIAL MANIFESTATIONS OF NUTRITIONAL PROBLEMS

- Weight loss or gain
- Loss of interest in food
- Anorexia
- Dry, rough, scaly skin
- Dull, dry, brittle, thin hair
- Loss of lean muscle mass
- Listlessness, apathy
- Poor muscle tone
- Constipation or diarrhea
- Irritability
- Fatigue and lack of energy
- Inflamed or bleeding gums

Potential Problems

1. Weight changes
2. Anorexia
3. Alteration in nutritional status due to drug therapy or complications of SLE

Nursing Interventions

Objective: Determine the causes of patient's altered nutritional status.

1. Conduct a physical assessment of patient, including weight, height, and percentage of body fat.
2. Assess patient's nutritional intake by asking her or him to keep a food diary.
3. Assess patient's current medications and doses.
4. Determine dietary and nutrient intake and vitamin/mineral supplement intake, food sensitivities (allergies may provoke a flare), food preferences, and experience with fad diets to "cure" lupus.
5. Assess patient for signs and symptoms of SLE-associated conditions, including osteoporosis, diabetes, and cardiovascular and kidney disease.
6. Monitor laboratory values such as hemoglobin, hematocrit, serum ferritin, serum iron, total cholesterol, HDL, LDL, VLDL, triglycerides, and plasma protein levels.

7. Assess patient for signs and symptoms of depression.
8. Assess patient's knowledge of nutrition and understanding of a healthful diet.
9. Assess patient's ability to purchase and prepare meals.
10. Assess patient's activity level.
11. Assess cultural, socioeconomic, and religious factors that may influence patient's diet.

Objective: Educate patient about healthful eating to prevent alteration in nutritional status.

1. Encourage patient to maintain a healthful diet, and discuss nutritional claims of "curing lupus," which are often misleading.
2. Provide patient with information on the basics of a well-balanced diet and its importance in a chronic disease such as lupus.
3. Instruct patient to take iron supplements only if iron stores are depleted.
4. Suggest vitamin and mineral supplementation, if necessary.
5. Refer patient to dietitian for assistance in dietary planning for serious conditions associated with SLE.

Note: For additional information, see the Patient Information Sheet (Chapter 7) on Nutrition and Lupus.

Chapter Five

MEDICATIONS USED TO TREAT LUPUS

edications are an important aspect of the management of many patients with SLE. An array of drug therapies is now available, which has increased the potential for effective treatment and excellent patient outcomes. Once a person has been diagnosed with lupus, a treatment plan will be developed by the doctor based on the person's age, health, symptoms, and lifestyle. It should be reevaluated regularly and revised as necessary to ensure it is as effective as possible. The goals for treating a patient with lupus include

- reducing tissue inflammation caused by the disease,
- suppressing immune system abnormalities that are responsible for tissue inflammation,
- preventing flares and treating them when they do occur, and
- minimizing complications.

PATIENTS AND PROVIDERS: WORKING TOGETHER

Lupus patients should work with their doctors to develop their medication treatment plan. Patients should thoroughly understand the reason for taking a drug, its action, dose, administration times, and common side effects. Pharmacists also can be a good resource for patients in helping them understand their medication treatment plan. If a patient experiences a problem believed to be related to a medication, the patient should notify her or his doctor immediately. It can be dangerous to suddenly stop taking some medications, and patients should not stop or change treatments without first talking to their doctor.

The array of drugs and the complexity of treatment plans can be overwhelming and confusing. Newly diagnosed patients and patients whose treatment plans have changed should be closely followed and have immediate access to a nurse or doctor if they are having problems with the prescribed medications. Most SLE patients do well on lupus medications and experience few side effects. Those who do experience negative side effects should not become discouraged, because alternative drugs are often available.

Health professionals should review drug treament plans with the lupus patient at each office visit to determine her or his understanding of and compliance with the plan. Questions should be encouraged and additional teaching done to reinforce or provide additional information as needed. It is important to note that lupus patients often require drugs for the treatment of conditions commonly seen with the disease. Examples of these types of medications include diuretics, antihypertensives, anticonvulsants, and antibiotics.

This chapter describes some of the main drugs used to treat SLE. The information presented is intended as a brief review and reference. Drug references and other medical and nursing texts provide more complete and detailed information regarding the use of each drug and associated nursing care responsibilities.

EDUCATING PATIENTS ABOUT LUPUS MEDICATIONS

Chapter 7 contains a Patient Information Sheet on each category of lupus medication covered here. The sheets can be used to teach lupus patients about the medications they may need to take. Each sheet contains general information about the category of drug, specific instructions on how and when to take the specific medication prescribed, information about possible side effects, and precautions.

Brand names included in this book are provided as examples only; their inclusion does not mean that these products are endorsed by NIH or any other Government agency. Also, if a particular brand name is not mentioned, this does not mean or imply that the product is unsatisfactory.

NONSTEROIDAL ANTI-INFLAMMATORY DRUGS (NSAIDS)

The NSAIDs comprise a large and chemically diverse group of drugs that possess analgesic, anti-inflammatory, and antipyretic properties. Pain and inflammation are common problems in patients with SLE, and NSAIDs are usually the drugs of choice for patients with mild SLE with little or no organ involvement. Patients with serious organ involvement may require more potent anti-inflammatory and immunosuppressive drugs.

Types of NSAIDs

There are as many as 70 NSAIDs on the market, and new ones are constantly becoming available. Some can be purchased as over-the-counter preparations, whereas larger doses of those drugs or other preparations are available only by prescription. For example, prescriptions are required for diclofenac sodium (Voltaren), indomethacin (Indocin), diflunisal (Dolobid), and nabumetone (Relafen).

Mechanism of Action and Use

The therapeutic effects of NSAIDs stem from their ability to inhibit the release of prostaglandins and leukotrienes, which are responsible for producing inflammation and pain. NSAIDs are very useful in treating joint pain and swelling and muscle pain. They may also be used to treat pleuritic chest pain. An NSAID may be the only drug needed to treat a mild flare; more active disease may require additional medications.

Although all NSAIDs appear to work in the same way, not every one has the same effect on every person. In addition, patients may do well on one NSAID for a period of time, then, for some unknown reason, derive no benefit from it. Switching the patient to a different NSAID should produce the desired effects. Patients should use only one NSAID at any given time.

Side/Adverse Effects

Gastrointestinal: Dyspepsia, heartburn, epigastric distress, and nausea; less frequently, vomiting, anorexia, abdominal pain, GI bleeding, and mucosal lesions. Misoprostol (Cytotec), a synthetic prostaglandin that inhibits gastric acid secretion, may be given to prevent GI intolerance. It prevents gastric ulcers and their associated GI bleeding in patients receiving NSAIDs.

Genitourinary: Fluid retention, reduction in creatinine clearance, and acute tubular necrosis with renal failure.

Hepatic: Acute reversible hepatotoxicity.

Cardiovascular: Hypertension and moderate to severe noncardiogenic pulmonary edema.

Hematologic: Altered hemostasis through effects on platelet function.

Other: Skin eruption, sensitivity reactions, tinnitus, and hearing loss.

Pregnancy and Lactation

NSAIDs should be avoided during the first trimester and just before delivery; they may be used cautiously at other times during pregnancy. NSAIDs appear in breast milk and should be used cautiously by breast-feeding mothers.

CONSIDERATIONS FOR HEALTH PROFESSIONALS

ASSESSMENT:

History: Allergy to salicylates, other NSAIDs, cardiovascular dysfunction, hypertension, peptic ulcer, GI bleeding or other bleeding disorders, impaired hepatic or renal function, pregnancy, and lactation

Laboratory data: Hepatic and renal studies, CBC, clotting times, urinalysis, serum electrolytes, and stool for guaiac

Physical: All body systems to determine baseline data and alterations in function, skin color, lesions, edema, hearing, orientation, reflexes, temperature, pulse, respirations, and blood pressure

EVALUATION:

Therapeutic response, including decreased inflammation and adverse effects

ADMINISTRATION:

With food or milk (to decrease gastric irritation)

TEACHING POINTS:

See Patient Information Sheet on NSAIDs.

ANTIMALARIALS

This group of drugs was first developed during World War II because quinine, the standard treatment for malaria, was in short supply. Investigators discovered antimalarials could also be used to treat the joint pain that occurs with rheumatoid arthritis. Subsequent use of antimalarials showed that they are effective in controlling lupus arthritis, skin rashes, mouth ulcers, fatigue, and fever. They have also been shown to be effective in the treatment of DLE. Antimalarials are not used to manage more serious, systemic forms of SLE that affect the organs. It may be weeks or months before the patient notices that these drugs are controlling disease symptoms.

Types of Antimalarials

The drugs most often prescribed are hydroxychloroquine sulfate (Plaquenil) and chloroquine (Aralen).

Mechanism of Action and Use

The anti-inflammatory action of these drugs is not well understood. In some patients who take antimalarials, the total daily dose of corticosteroids can be reduced. Antimalarials also affect platelets to reduce the risk of blood clots and lower plasma lipid levels.

Side/Adverse Effects

Central Nervous System: Headache, nervousness, irritability, dizziness, and muscle weakness.

Gastrointestinal: Nausea, vomiting, diarrhea, abdominal cramps, and loss of appetite.

Ophthalmologic: Visual disturbances and retinal changes manifested by blurring of vision and difficulty in focusing. A very serious potential side effect of antimalarial drugs is damage to the retina. Because of the relatively low doses used to treat SLE, the risk of retinal damage is small. However, patients should have a thorough eye examination before starting this treatment and every 6 months thereafter.

Dermatologic: Dryness, pruritus, alopecia, skin and mucosal pigmentation, skin eruptions, and exfoliative dermatitis.

Hematologic: Blood dyscrasia and hemolysis in patients with glucose 6-phosphate dehydrogenase (G6PD) deficiency.

Pregnancy

Antimalarials are considered to have a small risk of harming a fetus and should be discontinued in lupus patients who are attempting to become pregnant.

CONSIDERATIONS FOR HEALTH PROFESSIONALS

ASSESSMENT:

History: Known allergies to the prescribed drugs, psoriasis, retinal disease, hepatic disease, alcoholism, pregnancy, and lactation

Laboratory data: CBC, liver function tests, and G6PD deficiency

Physical: All body systems to determine baseline data and alterations in function, skin color and lesions, mucous membranes, hair, reflexes, muscle strength, auditory and ophthalmological screening, liver palpation, and abdominal examination

EVALUATION:

Therapeutic response and side effects

ADMINISTRATION:

Before or after meals at the same time each day to maintain drug levels

TEACHING POINTS:

See Patient Information Sheet on Antimalarials.

CORTICOSTEROIDS

Corticosteroids are hormones secreted by the cortex of the adrenal gland. SLE patients with symptoms that do not improve or who are not expected to respond to NSAIDs or antimalarials may be given a corticosteroid. Although corticosteroids have potentially serious side effects, they are highly effective in reducing inflammation, relieving muscle and joint pain and fatigue, and suppressing the immune system. They are also useful in controlling major organ involvement associated with SLE. These drugs are given in much higher doses than the body produces and act as potent therapeutic agents. The decision to use corticosteroids is highly individualized and is dependent upon the patient's condition.

Once the symptoms of lupus have responded to treatment, the dose is usually tapered until the lowest possible dose that controls disease activity is achieved. Patients must be monitored carefully during this time for flares or recurrence of joint and muscle pain, fever, and fatigue that can result when the dosage is lowered. Some patients may require corticosteroids only during active stages of the disease; those with severe disease or more serious organ involvement may need long-term treatment.

Treatment with corticosteroids must not be stopped suddenly if they have been taken for more than 4 weeks. Administration of corticosteroids causes the body's own production of adrenal hormones to slow down or stop, and adrenal insufficiency will result if the drug is stopped suddenly. Tapering the dose allows the body's adrenal glands to recover and resume production of the natural hormones. The longer a patient has been on corticosteroids, the more difficult it is to lower the dose or discontinue use of the drug.

Types of Corticosteroids

Prednisone (Orason, Meticorten, Deltasone, Cortan, Sterapred), a synthetic corticosteriod, is most often used to treat lupus. Others include hydrocortisone (Cortef, Hydrocortone), methlyprednisolone (Medrol), and dexamethasone (Decadron). Corticosteroids are available as a topical cream or ointment for skin rashes, as tablets, and as an injectable for intramuscular or intravenous administration.

Mechanism of Action and Use

The frequently prescribed corticosteroids are highly effective in reducing inflammation and suppressing the immune response. These drugs may be used to control exacerbation of symptoms and are used to control severe forms of the disease. The drug is usually

administered orally. During periods of serious illness, it may be administered intravenously; once the patient has been stabilized, oral administration should be resumed.

Side/Adverse Effects

Central Nervous System: Convulsions, headache, vertigo, mood swings, and psychosis.

Cardiovascular: Congestive heart failure (CHF) and hypertension.[1]

Endocrine: Cushing's syndrome, menstrual irregularities, and hyperglycemia.

Gastrointestinal: GI irritation, peptic ulcer, and weight gain.

Dermatologic: Thin skin, petechiae, ecchymoses, facial erythema, poor wound healing, hirsutism,[1] and urticaria.

Musculoskeletal: Muscle weakness, loss of muscle mass, and osteoporosis.[1]

Ophthalmologic: Increased intraocular pressure, glaucoma, exophthalmos, and cataracts.[1]

Other: Immunosuppression and increased susceptibility to infection.

[1] Long-term effects.

CONSIDERATIONS FOR HEALTH PROFESSIONALS

ASSESSMENT:

History: Hypersensitivity to corticosteroids, tuberculosis, infection, diabetes, glaucoma, seizure disorders, peptic ulcer, CHF, hypertension, and liver or kidney disease

Laboratory data: Electrolytes, serum glucose, WBC, cortisol level

Physical: All body systems to determine baseline data and alterations in function, weekly weight gain of >5 pounds, GI upset, decreased urinary output, increased edema, infection, temperature, pulse irregularities, increased blood pressure, and mental status changes (e.g., aggression or depression)

EVALUATION:

Therapeutic response, including decreased inflammation and adverse effects

ADMINISTRATION:

With food or milk (to decrease GI symptoms)

TEACHING POINTS:

See Patient Information Sheet on Corticosteroids.

Pregnancy and Lactation

Corticosteroids cross the placenta, but can be used cautiously during pregnancy. They also appear in breast milk; patients taking large doses should not breastfeed.

IMMUNOSUPPRESSIVES

Immunosuppressive agents are generally used to reduce rejection of transplanted organs. They are also used in serious, systemic cases of lupus in which major organs such as the kidneys are affected or in which there is severe muscle inflammation or intractable arthritis. Because of their steroid-sparing effect, immunosuppressives may also be used to reduce or sometimes eliminate the need for corticosteroids, thereby sparing the patient from undesirable side effects of corticosteroid therapy.

Immunosuppressives can have serious side effects. Patients need to understand, however, that side effects are dose dependent and are generally reversible by reducing the dose or stopping the medication.

Types of Immunosuppressives

A variety of immunosuppressive drugs is available to treat lupus. Although they have different mechanisms of action, each type functions to decrease or prevent an immune response. The immunosuppressives most frequently used with SLE patients are azathioprine (Imuran), cyclophosphamide (Cytoxan), methotrexate (Rheumatrex), and cyclosporine (Sundimmune, Neoral).

Mechanism of Action and Use

Drugs like azathioprine, methotrexate, and cyclosporine are referred to as antimetabolite agents. These drugs block metabolic steps within immune cells and then interfere with immune function. Cytotoxic drugs like cyclophosphamide work by targeting and damaging autoantibody-producing cells, thereby suppressing the hyperactive immune response and reducing disease activity.

Risks

There are many serious risks associated with the use of immunosuppressives. They include immunosuppression (resulting in increased susceptibility to infection), bone marrow suppression (resulting in decreased numbers of RBCs, WBCs, and platelets), and development of malignancies.

Side/Adverse Effects

Dermatologic: Alopecia (cyclophosphamide only).

Gastrointestinal: Nausea, vomiting, stomatitis, esophagitis, and hepatotoxicity.

Genitourinary: Hemorrhagic cystitis, hematuria, amenorrhea,[1] impotence,[1] and gonadal suppression (cyclophosphamide only).[2]

[1] Temporary or reversible once drug therapy is discontinued.

[2] Recovery of function after drug is discontinued is unpredictable.

CONSIDERATIONS FOR HEALTH PROFESSIONALS

ASSESSMENT:

History: Allergy to immunosuppressive drugs, infections, impaired hepatic or renal function, pregnancy, lactation, corticosteroid therapy, immunosuppression, and bone marrow suppression

Laboratory data: CBC, differential, platelet count, renal function studies, liver function tests, pulmonary function tests, chest X ray, and electrocardiogram (ECG)

Physical: All body systems to determine baseline data and alterations in function, temperature, pulse, respirations, weight, skin color, lesions, hair, and mucous membranes

EVALUATION:

Therapeutic response and adverse effects

ADMINISTRATION:

Orally or intravenously. **Precaution:** Drug administration protocols can vary. The nurse must work closely with the prescribing physician to safely administer the drug and to monitor the patient to minimize adverse effects and achieve expected outcomes

TEACHING POINTS:

See Patient Information Sheets on Azathioprine and Cyclophosphamide.

Hematologic: Thrombocytopenia, leukopenia, pancytopenia, anemia, and myelo-suppression.

Respiratory: Pulmonary fibrosis.[1]

Other: Increased risk of serious infections or malignancies.

Pregnancy and Lactation

Use of immunosuppressives presents definite risks to the fetus. Female patients should use contraceptive measures during treatment and for 12 weeks after ending azathioprine therapy. Azathioprine may pass into breast milk, and women using this drug should consult with their doctors before breastfeeding.

[1] With high doses.

Chapter Six

PSYCHOSOCIAL ASPECTS OF LUPUS

upus is a chronic disease that defies easy description. A diagnosis may not be made for a long time. Diagnostic difficulties coupled with the serious, chronic nature of the disease present the patient, family, and the medical team with many challenges. For lupus patients, psychosocial issues can be a major part of living with this illness.

Health professionals need to be aware of these issues to achieve excellent, comprehensive care of people with lupus. Because lupus is a chronic disease of flares and remissions, each exacerbation of the disease can raise new issues of coping for the patient. This chapter describes concerns typically raised by people confronting lupus and their families. Each of these concerns is an opportunity for the nurse or other health professional to provide education and to help people explore feelings and resolve problems.

SEEKING A DIAGNOSIS

It may take some time for a patient to be definitively diagnosed with lupus. During this time, patients may be confused or frustrated by the seeming inability of the doctors they visit to confirm the diagnosis. They may ask, "Why don't the doctors know?" Part of the difficulty, both for the patient and the doctor, rests in the fact that the diagnosis may seem to be hiding in a forest of confusing, vague, or changeable symptoms. A patient may express some of the following sentiments or frustrations:

"My symptoms are bizarre — they're here today and gone tomorrow."

"I can't put a handle on my symptoms. I'll have one today and a totally new one tomorrow."

"No one seems to believe me. My family thinks it is all in my head and they want me to see a psychiatrist. I am beginning to wonder if it is all in my head."

Before a diagnosis is made, many of a patient's primary needs are emotional. A lupus patient will, in all likelihood, be on intimate terms with her or his symptoms long before their cause is known. Realistically, she or he is the best authority on these symptoms. A patient may feel frustrated if, after describing symptoms, others do not respect her or his knowledge or do not share the conviction that something is wrong. If the doctor, family, or friends are unsupportive, the patient's fear, anger, and sense of isolation will only increase. These feelings add stress, which in turn can exacerbate the disease.

Health professionals can help ease these feelings by showing empathy during this difficult time and by reassuring the patient that the symptoms are real and merit serious attention. In addition, treating the patient as a whole person, and not just as a subject with a disease, can be immensely valuable in establishing a trusting relationship with the patient. Such a relationship will help the patient speak freely about symptoms or concerns that she or he may have been unwilling to discuss previously.

AFTER THE DIAGNOSIS

Patients will certainly experience a sense of relief once their condition is finally given a name and a tangible identity. At the same time, other emotions — anger, fear, depression, confusion, grief — may also surface. Patients may express some of the following sentiments:

"At last — a diagnosis! Now I know why I feel the way I do."

"Why me?"

"I'll never be able to make plans, because I don't know what tomorrow will bring."

"I feel guilty for having lupus and for all the trouble it's causing my family."

"Should I tell everyone or keep it quiet?"

"Will I lose my job?"

"I hate this disease. It's destroying my life and my family."

"I'm scared."

"Will I die soon?"

"Will I ever be able to have children?"

"How will we pay all these medical bills?"

After the diagnosis, some patients will have an insatiable desire for information about the disease; others may need to work through intense emotions before they can come to grips with their illness and begin to cope productively. The rapport that the health professional has established with the patient can now be used to provide the patient with information, resources, and an accepting atmosphere in which to adjust emotionally. This rapport can set a foundation of hope.

LIVING WITH LUPUS: KEY ISSUES

Every day, the patient living with lupus confronts a range of issues, from the emotional reactions of a spouse, children, or work colleagues to her or his own fluctuating physical and psychological state. The thoughtful and sympathetic health professional can contribute greatly by helping the patient see these issues objectively, understand that they will come and go over time, and develop effective coping skills.

Family Issues

One of the most important emotional issues that lupus patients grapple with is the ongoing and changeable reactions of those closest to them: parents, a spouse, or children. Understanding family dynamics can help the health professional work with the patient to develop positive coping strategies.

Parents

Parents of a lupus patient may react by smothering or — the other extreme — by not taking the disease seriously. Because lupus may be genetic, some parents may feel guilty for having "given" their child the disease. The patient may hear sentiments such as:

"Oh, my poor baby, let me take care of everything."

"Don't make such a big deal of it. Just shake it off."

"If it's genetic, I must have given it to you."

Spouse or Partner

A spouse or partner often experiences many of the same strong emotions the lupus patient does. Grief, fear, and anger are common emotions for spouses or partners as they deal with the patient's changing physical condition.

Well-established roles and responsibilities within the family may change, leading to confusion or conflict. These changes and feelings can affect the daily workings of the relationship, even threatening its foundation:

"I want you to get well. I want you to be the same as when I met you."

"I'm afraid you will die and leave me and the children."

"I'm confused. Our roles keep changing, and then changing back."

"It's hard doing my job and yours, too. I'll never learn how to do the work around the house well enough to suit you."

"I'm angry that you feel sick all the time and can't do what you used to. I feel guilty for being angry."

"Lupus has damaged our sex life. You're always too tired, and I'm afraid I'll hurt you."

"I need to mourn our losses, too."

Children

It is difficult for the children of a lupus patient to deal with the large and complex issues raised by having a sick parent. Some of these issues are tangible, whereas others are scary precisely because of their abstract, unknown nature. Because younger children have difficulty articulating their feelings and concerns, these emotions may go unnoticed or may be acted out in negative or disruptive behaviors. Older children with younger siblings may feel resentment as well as concern. Children's fears and feelings may emerge in statements such as:

"We never play anymore because you're always tired or sick."

"Is something terrible going to happen to you? Will you go away forever?"

"You've denied me my childhood. I don't want to be responsible for my little brother all the time."

The Patient's Own Physical and Emotional State

The physical repercussions of lupus, such as fatigue, weight gain, or an increased sensitivity to sunlight, can trigger intense emotional reactions. The following sentiments illustrate some common physical and emotional experiences of lupus patients:

Fatigue

"Nobody understands how it feels to be this tired."

"I feel like I'm trying to move through molasses. Even my bone marrow feels tired."

"No matter what I do or how much sleep I get, I still wake up tired."

"Between pain, stiffness, and fatigue, I feel like an old, old person."

Personal Appearance

"Why do I look so awful? I don't even look like myself."

"I don't want anyone to see the rash on my face."

"I used to have beautiful thick hair."

"I hate the fact that I eat so much, but this medicine makes me hungry all the time. I keep gaining weight."

Physical and Mental Abilities

"I've always loved sitting out in the sun. Now I can't anymore."

"I can't do the outdoor recreational things I used to do. I feel I'm letting my family down by not being able to play outdoors with them."

"Does anyone else have memory lapses? I'm afraid something terrible may happen because I'll forget something really important."

Depression

"I'm always on the verge of tears."

"I don't want to leave the house."

"I can never make plans, because I don't know what tomorrow will bring."

"Sometimes I think my family and friends would be better off without me."

The Future

"I've taken too much sick leave at work."

"Should I tell my boss I have lupus? I'm afraid I'll lose my job, and we really need the money."

"Will I pass the disease on to my children?"

"What will happen as I grow older? Am I going to die from this disease?

LIVING WITH LUPUS: DEVELOPING EFFECTIVE COPING SKILLS

Many lupus patients go through phases in which they feel that control over their life is slipping from their own hands into those of an unpredictable and unpleasant disease. This sense of powerlessness can occur not only during flares but also during periods of recuperation and remission. It forces the patient to choose between two options. This choice may be made many times during the course of the illness.

The first option is for the patient to submit to the disease and accept lupus and a lifestyle of illness as her or his identity. This choice may appear attractive to a newly diagnosed patient who is exhausted from the long battle of uncertainties related to lupus or to a long-term patient who is exhausted from fighting the disease. However, this option offers a life of self-pity, negativity, and significantly diminished horizons.

The second option is for patients to create a new identity based on reworked, realistic goals and expectations. Inherent in this second option is a sense of greater control, an improved self-image, and a positive and hopeful attitude. This option requires imagination, resilience, and determination and depends heavily on the existence of an adequate support network that can reinforce gains and buffer the occasional disappointment. This option offers true quality of life.

Perhaps the greatest gift that the health professional can offer patients is the opportunity to choose this second option. This gift can be given in several ways.

Help Patients Gain Control Over Feelings and Emotions

Patients must first assess their needs and the needs of those around them; evaluate their personal strengths, resources, and weaknesses; and develop effective communication strategies for dealing with family, friends, and the health care team. The health professional can assist the patient or suggest other professionals who can help. Many health professionals — for example, nurses, health educators, psychologists and psychiatrists, social workers, and occupational and physical therapists — are experienced in rheumatology and lupus. These professionals can educate family and friends about the needs and circumstances of lupus patients. The health professional can also encourage the patient to seek out other supportive mechanisms, such as:

- local support groups,
- educational and self-management programs offered by the Lupus Foundation of America, the Arthritis Foundation, and the SLE Foundation, and
- pen pals.

Help Patients Gain Control Over Their New Physical Limitations

Lupus patients need to accurately assess their pain and fatigue levels and understand how changes in these levels will affect their ability to work, play, and carry out ADL. Health professionals can help patients develop an effective self-management program that sets out achievable goals, realistically paces activities, and avoids overcommitments.

Help Patients Gain Control Medically

Doctors, nurses, and other professionals involved in lupus care can teach patients about many medical aspects of the disease, such as warning signs of an impending flare (the patient may have much to contribute to this discussion), medication use, possible side effects, and warning signals for contacting medical personnel. Nurses can also teach strategies for communicating with the health care team. This knowledge and these strategies will help the patient gain a sense of increased medical control over the disease. The Patient Information Sheets in Chapter 7 can help with this process.

Research advances are increasing the understanding of lupus and leading to improved treatments and medications. These advances and a growing body of practical experience in living with and controlling lupus point to a prognosis for lupus patients that is far brighter than it was even 20 years ago. Nurses and other health professionals who work on an ongoing basis with lupus patients can have a significant role in improving the emotional and psychological status of patients so that they can take advantage of this brighter tomorrow.

Chapter Seven

PATIENT INFORMATION

Caring for the lupus patient involves a number of critically important elements. Providing medical care as directed by the patient's doctor, monitoring the patient's physical status over time, and being sympathetic, understanding, and supportive are all involved. Educating patients and encouraging them to learn about their disease is another crucial element. The Patient Information Sheets in this chapter cover a range of topics about lupus and lupus medications and can help with this aspect of patient care. The topics are

- Living With Lupus,
- Preventing Fatigue Due to Lupus,
- Exercise and Lupus,
- Preventing a Lupus Flare,
- Serious Conditions Associated With Lupus,
- Joint Function and Lupus,
- Skin Care and Lupus,
- Fever and Lupus,
- Nutrition and Lupus,
- Sexuality and Lupus,
- Pregnancy and Lupus,
- Nonsteroidal Anti-Inflammatory Drugs (NSAIDs),
- Antimalarials,
- Corticosteroids,
- Azathioprine, and
- Cyclophosphamide.

TEACHING THE LUPUS PATIENT

The Patient Information Sheets provide a wealth of information, and are written in language that most patients will find easy to understand. Health professionals should hand them out to patients as appropriate during their discussions on specific issues related to lupus. The sheets can be photocopied directly from this guide, or they can be downloaded from the Web site of the National Institute of Arthritis and Musculoskeletal and Skin Diseases (*http://www.nih.gov/niams/*). A set of the Patient Information Sheets can also be ordered from the National Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse (NAMSIC) (see Chapter 8, *Resources and Bibliography*, for the NAMSIC address). The following points may help health professionals use them effectively.

Use the Sheets to Complement Existing Teaching Efforts

Over time, the doctor and other members of the health care team will probably discuss with a patient much of the information contained in these sheets. However, some patients may not absorb all the information given to them verbally. The Patient Information Sheets can be a useful backup. As the health professional talks through an issue, he or she may want to refer to or highlight specific sections of a sheet. This will help to reinforce the information and show the patient where to find it later.

Use the Sheets Selectively

The Patient Information Sheets cover a wide range of issues. Not all of them will be appropriate for each patient. For example, the sheet on Serious Conditions Associated With Lupus would not be appropriate for a patient with mild lupus. On the other hand, the sheets on Skin Care and Lupus and on Preventing Fatigue Due to Lupus may be particularly useful for that patient. One approach is to give the patient several of the more general Patient Information Sheets initially, then see which others are relevant as time goes on. When patients are first given a prescription for a lupus medication, they should also receive the Patient Information Sheet on that medication.

Use the Sheets in Tandem

The information contained in a number of the Patient Information Sheets is complementary, and it may be helpful to give the patient several sheets together. For example, the sheets on Exercise and Lupus and on Joint Function and Lupus would work well together, as would the sheets on Fever and Lupus and on NSAIDs or Corticosteroids. Several of the sheets that contain more general information, such as Living With Lupus or Preventing a Lupus Flare, would be a good complement to many of the sheets dealing with more specific topics.

Chapter Eight

RESOURCES AND BIBLIOGRAPHY

urses and other health professionals may find the following organizations and written materials useful as sources of further information about lupus and patient care for the disease.

RESOURCES

National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS)

The mission of the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS), a part of the National Institutes of Health (NIH), is to support research into the causes, treatment, and prevention of arthritis and musculoskeletal and skin diseases, the training of basic and clinical scientists to carry out this research, and the dissemination of information on research progress in these diseases. The National Institute of Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse is a public service sponsored by the NIAMS that provides health information and information sources. Additional information can be found on the NIAMS Web site at www.niams.nih.gov.

1 AMS Circle
Bethesda, MD 20892–3675
Phone: (301) 495–4484 or
(877) 22-NIAMS (226-
4267) (free of charge)
TTY: (301) 565–2966
http://www.niams.nih.gov

NIAMS participates in the Combined Health Information Database (CHID), a large federally supported database of information on a broad range of diseases and related health issues, including lupus. CHID is on the Web at *http://chid.nih.gov/*.

National Institute of Nursing Research

The National Institute of Nursing Research (NINR), a component of NIH, supports research and research training in universities, hospitals, research centers, and at NIH in areas related to health promotion and disease prevention, managing the symptoms and disabilities of illness, and improving the environment in which patient care is delivered. Chronic diseases, such as lupus and arthritis, are among the six major areas of emphasis for the Institute. For more information on the Institute and its programs, contact the NINR Office of Science Policy and Information at:

31 Center Drive
MSC 2178
Building 31, Room 5B10
Bethesda, MD 20892
Phone: (301) 496–0207
http://www.nih.gov/ninr/

Lupus Foundation of America

The Lupus Foundation of America (LFA) is the main voluntary organization devoted to lupus. LFA assists local chapters in providing services, including education, referrals, and support groups, to people with lupus; works to educate the public about lupus; and supports lupus research. For more information, contact LFA at:

1300 Piccard Drive, Suite 200
Rockville, MD 20850–4303
Phone: (301) 670–9292 or
(800) 558–0121
http://www.lupus.org/

SLE Foundation, Inc.

The SLE Foundation supports and encourages medical research to find the cause and cure of lupus, and improve its diagnosis and treatment. It also provides a wide variety of services to help lupus patients and their families. In addition, this voluntary organization conducts a broad-based public education program to raise awareness of lupus, and increase understanding of this serious chronic autoimmune disease.
For more information, contact the SLE Foundation at:

149 Madison Ave., Suite 205
New York, NY 10016
Phone: (212) 685–4118
http://www.lupusny.org/

Association of Rheumatology Health Professionals, American College of Rheumatology

The American College of Rheumatology (ACR) is an organization of doctors and associated health professionals who specialize in arthritis and related diseases of the bones, joints, and muscles. The Association of Rheumatology Health Professionals (ARHP), a division of ACR, aims to enhance the knowledge and skills of rheumatology health professionals and to promote their involvement in rheumatology research, education, and quality patient care. The Association also works to advance and promote basic and continuing education in rheumatology for health professionals who provide care to people with rheumatic diseases. For more information, contact ARHP at:

1800 Century Place
Suite 250
Atlanta, GA 30345–4300
Phone: (404) 633–3777
http://www.rheumatology.org/

Arthritis Foundation

The Arthritis Foundation is the major voluntary organization devoted to supporting arthritis research and providing educational and other services to individuals with arthritis. It publishes free pamphlets and a magazine for members on all types of arthritis. It also provides up-to-date information on research and treatment, nutrition, alternative therapies, and self-management strategies. Chapters nationwide offer exercise programs, classes, support groups, physician referral services, and free literature. For more information, call your local chapter, listed in the white pages of the phone book, or contact the Arthritis Foundation at:

1330 W. Peachtree Street
Atlanta, GA 30309
Phone: (800) 283–7800
http://www.arthritis.org/

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ADDENDUM

The U.S. Food and Drug Administration (FDA) reviews and approves new drugs to treat lupus. For information on new drug treatments, please contact the FDA:

US Food and Drug Administration
Center for Drug Evaluation and Research
5600 Fishers Lane
HFD 240, Room 12B31
Rockville, MD 20857
Phone: (301) 827-4570
E-mail: druginfo@cder/fda.gov
http://www.fda.gov/oc/oha/default.htm

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Patient Information Sheet

Living With Lupus

You have recently been diagnosed with a disease known as systemic lupus erythematosus (SLE). It has probably taken time to arrive at this diagnosis. Now that you know, you may feel relieved but also overwhelmed. You probably have a lot of questions about lupus.

You may have a mild or a more serious form, but no matter how severe your lupus is, you will need close medical supervision. You may also need to make lifestyle changes to keep your disease under control and feel as well as possible. At the beginning, you may feel some of these emotions:

- anger or depression over the change in your health,
- uncertainty about what to tell family, friends, or coworkers,
- guilt for having lupus and the burden it may cause your family,
- fear that you may lose your job if you can no longer work regularly, and
- fear that you may die.

These are all normal feelings, and you are not alone in having them. You should give yourself time to adjust to your illness. This may or may not be easy for you. Discuss your feelings and concerns with your doctor and nurse and with your family and friends. Sometimes, talking with other people who have lupus is helpful. If you are having a hard time adjusting to your diagnosis, consider seeking the help of a counselor.

Many physical and emotional issues confront people with lupus, both in the beginning and throughout the course of their disease. The most common issues include the following.

Fatigue: Fatigue is a chronic problem that is usually accompanied by joint pain and stiffness. It can affect many aspects of your daily life.

Changes in personal appearance: You may experience changes in your personal appearance. Discoid lupus (a form of lupus) may cause sores, blotches, or scarring on the face, arms, shoulders, neck, or back. The medications for lupus can also sometimes change your appearance. For example, corticosteroids can cause weight gain, excessive hair growth, or swelling. Some drugs may cause hair loss. These changes in the way you look can be emotionally challenging to deal with.

Continued on next page

CARING FOR YOURSELF

- ▲ Learn as much about lupus as possible.
- ▲ Understand that you will experience a variety of emotions, particularly when you are first diagnosed and adjusting to the fact that you have lupus.
- ▲ Adopt a positive attitude.
- ▲ Evaluate your personal strengths and resources, such as family, friends, coworkers, and community ties.
- ▲ Determine what your needs are, then make a plan to address them.
- ▲ Don't be afraid to set goals for yourself, but be flexible.
- ▲ Learn how to manage the physical aspects of your disease and the effects they have on other areas of your life.
- ▲ Learn to deal with stressful situations, because stress and anxiety can make your lupus symptoms worse.
- ▲ Learn to talk with your health care team, family, friends, and coworkers about lupus and the effect it has on your life.
- ▲ Don't be afraid to seek help for yourself or your family.

REMEMBER: Living well with lupus is possible. It is important that you take control of your illness and not allow it to take control of you. Adopting a positive attitude and striving to be happy can make a big difference in the quality of your life and that of your family and friends.

Changes in physical ability: Many people with lupus feel isolated because their fatigue and need to rest keep them from maintaining normal work and social schedules. You may feel frustrated if you can't participate in outdoor activities with family or friends because of sensitivity to the sun. There will be times when you may feel it is easier to stay home than to make plans and later cancel them because you are too tired or not feeling well.

Psychological effects of corticosteroids: Corticosteroids are used to treat many of the symptoms of lupus that result from inflammation. Their use can cause anxiety, mood changes, forgetfulness, depression, personality changes, and other psychological problems. You need to know about the possible side effects of these drugs while you are taking them. It is also important that your family and friends understand the effects of these drugs so that they can be supportive if you should experience any side effects.

Depression: You may feel sad or depressed at times in your struggle to control lupus or because of the medications you take. Good communication with your doctor and health care team, as well as with your family and friends, is important in helping you cope with these feelings.

Concern for the future: Because the future and course of your disease are unknown, planning for your job, your family, and life in general can be difficult at times.

Family concerns: Like you, your family can be overwhelmed about your diagnosis and may have a difficult time understanding and adapting to your disease. They may feel confused, helpless, and afraid. Because of your physical limitations, traditional roles and responsibilities within the family may need to change. It is important that everyone talk openly and honestly with each other. It is also important that your family learns about your disease so they can better understand your physical and emotional condition and the changes in your family that may result.

ADDITIONAL NOTES

From *LUPUS: A Patient Care Guide for Nurses and Other Health Professionals*
National Institute of Arthritis and Musculoskeletal and Skin Diseases/National Institutes of Health

Patient Information Sheet

Preventing Fatigue Due to Lupus

Fatigue is a very common complaint of all people with systemic lupus erythematosus (SLE), even when no other symptoms of active disease are present. The fatigue of lupus isn't just being tired. You may feel an extreme fatigue that interferes with many aspects of your daily life. You may find that you are unable to participate in your normal pattern of daily activities, such as working, caring for your family and home, or participating in social activities. The exact cause of this fatigue is not known.

Your doctor and nurse will probably ask you about your lifestyle and patterns of daily living and activity. They will also evaluate your overall fitness, health, nutrition, and ability to handle stress. Your doctor or nurse will then be able to advise you about how your fatigue can be reduced. It is important to remember that getting enough rest, maintaining physical fitness, and keeping stress under control are absolutely necessary for anyone with lupus.

Changes in your lifestyle and patterns of daily living and activity may not be easy to accept. In addition, the changes necessary for you to cope with your disease today may be different from the changes you may have to make later as your disease changes. A positive attitude and a well-thought-out, but flexible, plan of action will increase the chances that you can make these changes successfully.

ADDITIONAL NOTES

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CARING FOR YOURSELF

- Get enough sleep. You may be able to get by on 8 hours a night, or you may need more.
- Plan for additional rest periods throughout the day, as needed. Do not exhaust yourself.
- Getting enough rest does not mean no activity at all. A well-designed exercise program is important to maintaining strength, endurance, and overall fitness.
- Every week, make a simple plan of your work and activities. The plan can help you organize the events of your life and ensure that you have a good balance of rest and activity.
- Each day, review your plan and decide if you are physically up to the activities for that day. Be flexible; if you don't have the strength to do an activity today, do it another time.
- Don't try to complete a large task or project all at one time; divide it into several steps.
- Eat a well-balanced diet.
- Dealing with stressful issues and problems takes a lot of energy. If you feel stressed out, talk with your doctor or nurse. They may be able to provide you with help for your problem or direct you to someone else who can.

From *LUPUS: A Patient Care Guide for Nurses and Other Health Professionals*
National Institute of Arthritis and Musculoskeletal and Skin Diseases/National Institutes of Health

Patient Information Sheet

Exercise and Lupus

Because lupus causes joint pain and inflammation, muscle pain, and fatigue, the very thought of exercising can be a challenge. In addition, because lupus is a disease that requires a large amount of rest, you might wonder why exercise is so important. Although rest is important in managing fatigue, too much rest can be harmful to muscles, bones, joints, and overall fitness. Keeping fit through an exercise program planned just for you can help you feel better, both mentally and physically. There are many types of exercises that are appropriate for lupus patients, such as swimming and walking. Regular exercise will

- increase your muscle strength,
- help prevent your joints from getting stiff,
- help prevent osteoporosis,
- help keep your weight under control,
- improve your cardiovascular health, and
- help reduce stress.

GETTING STARTED CAN BE THE HARDEST PART

- Check with your doctor before you start any type of exercise program. He or she can evaluate your overall condition and fitness and recommend a type and level of activity that are right for you.
- Try to find someone to exercise with; it can be a lot more fun.
- Start slowly. Chart your progress so you can see and take pride in your accomplishment.
- Change your exercise activities depending on how you feel. If you're not up to it on one day, that's okay. Try to go back to your program the next day.

For a healthy person, achieving physical fitness can be a lot of hard work. For a person with lupus, such an achievement is really something to be proud of!

ADDITIONAL NOTES

From *LUPUS: A Patient Care Guide for Nurses and Other Health Professionals*
National Institute of Arthritis and Musculoskeletal and Skin Diseases/National Institutes of Health

Patient Information Sheet

Preventing a Lupus Flare

Your doctor has put together a treatment plan that is designed specifically for you and your lupus. This probably includes physical and emotional rest, aggressive treatment of infections, good nutrition, and avoidance of direct sunlight and other sources of ultraviolet light. Your doctor may have also prescribed medications to control disease symptoms and other health problems that you might have. One of the most important ways you can help yourself is to understand your treatment plan and the things you need to do to keep your disease under control.

Sometimes, despite the treatment plan and your efforts, you may experience a lupus flare. A flare is a worsening of symptoms that signals increased disease activity. A variety of factors can cause a flare, and you should contact your doctor immediately if you suspect a flare is developing. The doctor will evaluate your condition and take steps to control the seriousness of the flare. He or she will also reevaluate your overall treatment plan and make any needed changes.

WARNING SIGNS OF A FLARE

- Increased fatigue
- A new or higher fever
- Increased pain
- Development or worsening of a rash
- Upset stomach
- Headache or dizziness
- Development of symptoms you haven't had before

WHAT TRIGGERS A FLARE?

A flare can be triggered by one factor or a combination of factors. The most common are

- overwork or not enough rest;
- stress or an emotional crisis;
- exposure to sunlight or other sources of ultraviolet light;

CARING FOR YOURSELF

- ▲ Learn to recognize the warning signals of a flare and tell your doctor about them.
- ▲ Maintain your physical health. Be sure to visit your doctor regularly, even if you are feeling well. Schedule regular dental, eye, and gynecological exams.
- ▲ Get enough sleep and rest. Be flexible with your schedule of daily activities.
- ▲ Try to limit your stress. Because this may be hard to do at times, consider developing a plan for dealing with potentially stressful situations. Develop a support system that includes family, friends, medical or nursing professionals, community organizations, and support groups. Remember, it helps to talk to someone when you're feeling stressed.
- ▲ Participate in a well-planned exercise program to help you maintain physical fitness and reduce stress.
- ▲ Eat a healthy diet.
- ▲ Limit your exposure to the sun and other sources of ultraviolet light, such as fluorescent or halogen lights.
- ▲ Tell your doctor right away about any injury, illness, or infection or if you do not feel well in any way.
- ▲ Delay elective surgery (including dental surgery and teeth pulling) until your lupus is under control or in remission.

Continued on next page

WHAT TRIGGERS A FLARE (*CONTINUED*)

- infection;
- injuries or surgery;
- pregnancy or the time right after the baby's birth (the postpartum period);
- sudden stopping of medications for lupus;
- sensitivities or allergies to items that you put on your skin, such as hair dye, hair permanent solution, makeup, and skin creams;
- certain prescription drugs;
- over-the-counter medications, such as cough syrup or laxatives; and
- immunizations.

ADDITIONAL NOTES

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- ▲ Lupus may cause problems for a pregnant woman and her baby. As a result, women with lupus should carefully plan any pregnancy. Do not stop using your method of birth control until you have discussed the possibility of pregnancy with your doctor and he or she has determined that you are healthy enough to become pregnant.
- ▲ Talk with your doctor before you stop taking any prescribed medications.
- ▲ Check with your doctor or nurse before taking any over-the-counter medications.
- ▲ Be careful when trying any over-the-counter preparations used on your skin or scalp. First, determine whether you have a sensitivity or an allergy to it. Put a small amount of the preparation on the inside of your forearm or on the back of your ear. If any redness, rash, raised areas, itching, or pain develops, do not use the preparation.
- ▲ Be aware that certain prescription drugs may trigger a flare. Tell any doctor, nurse, or health care professional you visit that you have lupus. Also tell your lupus doctor or nurse if any new medications have been prescribed for you.
- ▲ Be sure to check with your lupus doctor before receiving any immunization. Routine immunizations, including those for the flu and pneumonia, are an important part of maintaining your health, and you should get them if your doctor approves.

From *LUPUS: A Patient Care Guide for Nurses and Other Health Professionals*
National Institute of Arthritis and Musculoskeletal and Skin Diseases/National Institutes of Health

Serious Conditions Associated With Lupus

Although lupus can be well controlled in many people, serious medical conditions caused by or associated with the disease can still occur. It is important that you know about these conditions and how they may make you feel so that you can call your doctor right away. The sooner a problem is detected and evaluated, the sooner it can be treated to prevent or reduce damage to your body's organs.

Kidney disease: Many people with lupus develop some form of mild kidney disease. Others, however, develop kidney disease serious enough to lead to kidney failure. Warning signs include

- swelling around your ankles, hands, and eyes;
- increased fatigue or tiredness, especially if you have not altered your rest and activity patterns; and
- increased need to urinate at night.

Pericarditis: Pericarditis is an inflammation of the thin sac that surrounds the heart. Warning signs include

- chest pain,
- shortness of breath, and
- new or higher-than-usual fever.

Myocarditis: Myocarditis is an inflammation of the heart muscle. Warning signs include

- chest pain,
- shortness of breath, and
- new or higher-than-usual fever.

Atherosclerosis: This is a condition in which fatty deposits build up on the inside of arteries. These deposits can reduce or block blood flow. A blockage or reduced blood flow through an artery that supplies the heart can cause a heart attack to occur. Warning signs include

- burning, choking, squeezing, or pressing chest pain felt in the center of the chest that may radiate to the left shoulder and arm (anginal pain); it can last up to 5 minutes and will become much less intense or go away completely if you rest;
- crushing, prolonged chest pain that is not relieved by rest;
- shortness of breath;
- unrelieved indigestion; and
- a weak or faint feeling.

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ADDITIONAL NOTES

Pleuritis: Pleuritis is an inflammation of the lining of the lung. Warning signs include

- shortness of breath, and
- chest pain, especially when taking a deep breath.

Central nervous system (CNS) disease: CNS disease covers a variety of problems that may or may not be related to lupus. Problems can include seizures, memory loss, headache, confusion, hearing and visual changes, muscle weakness, depression, and emotional disturbances. Because many of these problems can be related to use of medications or indicate other conditions, it is often difficult to make a definite diagnosis of CNS disease. Warning signs include

- severe or chronic headaches;
- seizures;
- periods of forgetfulness, restlessness, or confusion;
- new or increased hearing and vision problems;
- bizarre or erratic changes in behavior;
- mood swings; and
- signs of a stroke, including weakness or numbness in the arms, legs, face, or down one side of the body; a change in speech; confusion; or severe headaches.

Depression: With depression, people may feel helpless, hopeless, or overwhelmed. They may find it difficult to get through the day. Depression can occur as a result of lupus or be caused by the drugs used to treat it, especially corticosteroids. Warning signs include

- depressed mood;
- significant weight loss or gain;
- trouble sleeping or sleeping too much;
- extreme tiredness and lack of energy;
- decreased concentration or an inability to make a decision;
- feelings of being overwhelmed and unable to carry out simple tasks, such as personal hygiene, housework, or childcare;
- feelings of hopelessness about various aspects of life;
- unusual anger or irritability; and
- recurrent thoughts of death and suicide.

Osteonecrosis: This is a condition that usually affects the hip joint, but may occur in other joints such as the knees, ankles, or shoulders. Blood supply to the hip is reduced and, over time, leads to severe degenerative arthritis. Osteonecrosis is considered to be a side effect of corticosteroid therapy and not a manifestation of SLE itself. Warning signs include

- sharp pain in the groin or buttocks that may radiate down the back of the leg,
- decreased exercise tolerance,
- stiffness of the hips, and
- increased pain and difficulty in walking after exercise.

Pancreatitis: In pancreatitis, the pancreas (an organ involved in digestion and in producing hormones that regulate blood sugar levels) becomes inflamed. It is a very serious problem that must be treated immediately. Warning signs include

- sharp, intense pain at the level of the belly button that radiates around to the back,
- nausea and vomiting, and
- new or higher-than-usual fever.

Acute abdomen: This is a condition that describes the sudden onset of abdominal pain. A variety of serious problems can cause this condition. You should see your doctor immediately if you develop acute abdomen. Warning signs include

- abdominal pain that may be severe and radiate throughout the abdominal area;
- nausea, vomiting, or loss of appetite;
- change in usual bowel movements; and
- vomiting blood or blood in the stool.

Vision problems: Changes in vision can be a result of lupus or because of the corticosteroids and antimalarials used to treat lupus. Problems can include inflammation of the eye, glaucoma, cataracts, general changes in vision, and blocked tear ducts. On very rare occasions, blindness can result. Warning signs include

- development of a rash over the eyelids;
- mucus discharge from the eye;
- blurred vision;
- sensitivity to light;
- headaches;
- a sore, red eye;
- lack of tears, and eyes that hurt and are dry; and
- episodes of flashing lights and partial blindness.

From *LUPUS: A Patient Care Guide for Nurses and Other Health Professionals*
National Institute of Arthritis and Musculoskeletal and Skin Diseases/National Institutes of Health

Joint Function and Lupus

Joint pain or arthritis is experienced by 95% of people with lupus at some time during the course of their disease. In fact, joint pain is usually the first symptom of lupus. Unlike rheumatoid arthritis, the arthritis of lupus tends to be temporary. It is also less damaging to the joints. The joints most commonly involved are those of the fingers, wrists, and knees. Elbows, ankles, and shoulders are not affected as often. When a particular joint is affected on one side of the body, the same joint on the other side of the body is usually affected as well.

Arthralgia: Arthralgia means "joint pain." Morning stiffness, swelling, or heat in the joints can also occur.

Myalgia or myositis: Myalgia means "pain in the muscles"; myositis means "inflammation of the muscle." These may include overall muscle pain and tenderness, especially in the upper arms and upper legs. They are common in 40–80% of people with lupus, especially during a flare.

Other joint complications: Several types of joint complications occur rarely in lupus. They include osteonecrosis (damage to the hip joint that leads to severe arthritis), development of nodules in the small joints of the hands, tendinitis, tendon rupture, and carpal tunnel syndrome. Your doctor or nurse can give you more information about these problems.

TAKING CARE OF YOUR JOINTS

If you have joint or muscle problems, the first goal is to keep pain at a tolerable level. You can do this in several ways:

- Apply heat or cold to the affected joints.
- Support the affected joints with pillows, blankets, or splints (if ordered by your doctor).
- Rest the affected joints as much as possible and keep them elevated to help reduce swelling.
- Follow your doctor's plan for managing pain and using anti-inflammation medication.

ADDITIONAL NOTES

Continued on next page

Your second goal is to maintain joint function and increase muscle strength. You can do this by using the following techniques:

- Take warm showers or baths to lessen stiffness.
- Don't put any weight on an acutely inflamed joint. Sit or lie down. Avoid strenuous activity and avoid any activity that causes increased pain, swelling, tenderness, or heat to the affected joint.
- Ask a physical therapist or trained family member or friend to gently move the inflamed joint in all the directions it can be moved (this is called passive range of motion [ROM]).This will help prevent stiffness. Your doctor can let you know when and how often this should be done.
- Gently move the affected joint yourself when the acute inflammation is over.
- Talk with your doctor or nurse about physical or occupational therapy if you are having trouble regaining joint strength and motion or if activities of daily living (cooking, cleaning, bathing, etc.) are still difficult.
- Hire a housekeeper or someone to help care for yourself or your children until you feel better.

When you are feeling better and your physical condition has improved, your doctor will probably recommend an exercise program tailored to your needs. Although rest and protecting joint function are extremely important, exercise is also necessary to keep muscles, bones, joints, and tendons strong and healthy. A well-planned exercise program combined with other aspects of your care will help you maintain joint function and improve your overall fitness.

ADDITIONAL NOTES

From *LUPUS: A Patient Care Guide for Nurses and Other Health Professionals*
National Institute of Arthritis and Musculoskeletal and Skin Diseases/National Institutes of Health

Patient Information Sheet

Skin Care and Lupus

Skin problems are very common in people with lupus. Some skin rashes and sores (also called lesions or ulcers) are very specific to lupus, while others can occur in other diseases as well. A sensitivity to and too much exposure to the ultraviolet (UV) rays of sun and some types of artificial light are responsible for aggravating some rashes and lesions. Many types of skin conditions are common in lupus.

Butterfly rash: This rash over the nose and cheeks can range from a faint blush to a rash that is very severe, with scaling. It is very sensitive to light and appears to gets worse when skin is exposed to sun or certain types of artificial light. The rash may be permanent or may come and go.

Discoid lesions: These scarring, coin-shaped lesions are seen on areas of the skin that have been exposed to UV light. They may also occur on the scalp and produce a scarring, localized baldness that is permanent.

Subacute cutaneous lesions: These nonscarring, red, coin-shaped lesions are very sensitive to UV light. They can appear scaly and can mimic the lesions seen in psoriasis. They may occur only on the face or cover large areas of the body.

Mucous membrane lesions: Mouth ulcers are sometimes seen in lupus patients. Nose and vaginal ulcers may also occur. These lesions are usually painless.

Hair loss: In addition to losing hair because of discoid lesions, some lupus patients may develop a temporary, generalized hair loss followed by the growth of new hair. Hair loss may also be caused by infection or by use of corticosteroids or other lupus medications. A severe lupus flare could result in defective hair growth, causing the hair to be fragile and break easily.

Vasculitis: This is a condition in which the blood vessels become inflamed. Very small blood vessels can break and cause bleeding into the tissues, resulting in tiny, reddish-purple spots on the skin known as petechiae (pe-teke-ee-ah). Larger spots are called purpura and may look like a bruise. Vasculitis can also cause blood clots to form, skin

CARING FOR YOURSELF

- ▲ Reduce your exposure to the sun and to some sources of artificial light (especially fluorescent and halogen bulbs). The skin of people with lupus is very sensitive to the UV light that comes from these sources.
- ▲ Limit outdoor activity between the hours of 10 a.m. and 4 p.m. This may mean a big change in your lifestyle if you work or play outdoors a lot.
- ▲ Wear a sunscreen on exposed areas of skin. It should have a sun protection factor (SPF) of 15 or higher. Be sure that the sunscreen protects against both UVB and UVA rays.
- ▲ Wear sunscreen all year round and on cloudy days as well as on sunny days. Also wear it indoors if you spend a lot of time in a room with many windows (glass does not filter out UV rays).
- ▲ Wear protective clothing, such as hats with wide brims and clothing made of tightly woven material. Thin, loosely woven material allows UV light to penetrate to the skin.
- ▲ Be aware of fluorescent light and halogen lamps. They can be found in many places and include floor lamps, overhead lights, photo-copiers, and slide projectors. Sunscreen and protective clothing can help.
- ▲ Tell your doctor immediately if any rash or sore appears or gets worse.

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ulcers to develop, and small black areas to appear around fingers and toenails. These black areas are a sign of serious tissue damage. If they begin to develop, see your doctor immediately.

Raynaud's phenomenon: This is a condition in which the blood vessels of the fingers and toes react in an extreme way to cold or stress. They suddenly get very narrow (vasoconstrict). This decreases the blood supply going through the vessel. As a result, the fingers and toes become cold and can become pale or bluish. Pain or tingling can occur when the hands and feet warm up and circulation returns to normal.

Drug-induced skin changes: Some drugs used to treat lupus, such as corticosteroids, immunosuppressives, and antineoplastics, can affect the skin. Your doctor or nurse will review these side effects with you if one of these drugs has been prescribed.

ADDITIONAL NOTES

- If your doctor prescribes a medication for your skin condition, be sure to take it as directed.
- Try rinsing your mouth with salt water and eating soft foods if you have mouth ulcers. A number of other treatments and preparations are available to treat mouth ulcers as well as those in the nose and vagina.
- Avoid preparations or medications you know will make your skin condition worse. These might include hair dyes, skin creams, certain drugs that can make you more sensitive to the sun (for example, tetracyclines or diuretics), and things you are allergic to.
- It's okay to wear makeup, but try hypoallergenic brands. A brand that also includes UV protection would be good to use.
- If you have Raynaud's phenomenon, dress warmly in cold weather. Pay particular attention to keeping your hands and feet warm. Keeping your home warm will also help prevent an attack. Avoid smoking, caffeine, and stress — all of these can contribute to Raynaud's phenomenon.
- If you have trouble maintaining a positive attitude about your appearance or your lupus, call your doctor or nurse to discuss your feelings and concerns.

From *LUPUS: A Patient Care Guide for Nurses and Other Health Professionals*
National Institute of Arthritis and Musculoskeletal and Skin Diseases/National Institutes of Health

Patient Information Sheet

Fever and Lupus

Fever is often a part of lupus. For some people with lupus, an intermittent (coming and going) or continuous low-grade fever may be normal. Other people, especially those on large doses of aspirin, nonsteroidal anti-inflammatory drugs (NSAIDs), or corticosteroids, may not have fever at all because these drugs may mask a fever.

If you have lupus, you may be more vulnerable to certain infections than are other people without lupus. In addition, you may be more prone to infection if you are taking any immunosuppressive drugs for your lupus. Be alert to a temperature that is new or higher than normal for you, because it could be a sign of a developing infection or a lupus flare.

ADDITIONAL NOTES

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CARING FOR YOURSELF

- Take your temperature at least once a day (or more often if needed) to determine what a "normal" temperature is for you.
- Take your temperature and watch for a fever any time you feel chills or do not feel well.
- Call your doctor immediately if you have a new or higher-than-normal temperature.
- Even if you don't have a fever, don't hesitate to call your doctor if you do not feel well in any way, particularly if you are taking aspirin, NSAIDs, or a corticosteroid. Signs of infection other than a fever include unusual pain, cramping or swelling, a headache with neck stiffness, cold or flu symptoms, trouble breathing, nausea, vomiting, diarrhea, or changes in urine or stool.
- Talk to your doctor about immunization against pneumococcal pneumonia and the flu.
- Practice good personal hygiene.
- Avoid large crowds and people who are sick.

From *LUPUS: A Patient Care Guide for Nurses and Other Health Professionals*
National Institute of Arthritis and Musculoskeletal and Skin Diseases/National Institutes of Health

Nutrition and Lupus

Good nutrition is an important part of the overall treatment plan for your lupus. A well-balanced diet provides the necessary fuel for your body to carry on its normal functions. Although there are no specific dietary guidelines for people with lupus, there are some nutrition issues that you should know about. If any of these issues become a problem for you, talk with your doctor or nurse. They will be able to provide you with additional information and can refer you to a registered dietitian if necessary.

Weight loss or poor appetite: Weight loss over the previous year is commonly reported by people who are newly diagnosed with lupus. Weight loss and poor appetite can be caused by the illness itself or by some medications that may cause stomach upset or mouth sores (also called mouth ulcers).

Your doctor or nurse will assess your weight loss and other related problems and suggest changes in your diet to be sure that you are eating right and have no further weight loss.

Weight gain: Weight gain may be a problem for people who take corticosteroids. These drugs often increase a person's appetite, and, unless you are careful, unwanted weight gain will occur.

Your doctor or nurse will assess your diet and other related problems and can suggest a program to help you control your weight and lose any unwanted pounds. The program will probably include a low-fat diet, exercise, and behavior modification. A registered dietitian can help you evaluate your food likes and dislikes and eating patterns and can design a diet specifically for your needs and lifestyle.

Difficulty taking medications: Several medications can cause gastrointestinal (GI) disturbances, such as heartburn, upset stomach, nausea, vomiting, or painful mouth ulcers.

If you are having GI problems, tell your doctor or nurse immediately. Because many of these problems are related to how and when a medication is taken, the dose or schedule can sometimes be changed to reduce or stop the unpleasant side effects. In some cases, the

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ADDITIONAL NOTES

doctor may change the drug. Many medications can be taken with food, which helps reduce side effects. If you have mouth ulcers, liquid forms of the drugs you are taking may be available. In addition, anesthetics you can use in your mouth can decrease the pain of mouth ulcers and make swallowing easier.

Osteoporosis: Osteoporosis is a condition in which the bones of the body become less dense and break easily. Although this condition often affects older, postmenopausal women, it can also affect anyone who takes corticosteroids for a long period of time.

Your doctor or nurse will review your medical history, treatment plan, diet, and any risk factors you may have. Measurements of your bone density may also be taken. Recommendations to prevent or reduce the problem will probably include a diet high in calcium (1,000–1,500 mg/day) and vitamin D (100–500 mg/day) and an exercise plan that is appropriate for you. Calcium supplements may be prescribed by your doctor if the calcium in your diet is not enough.

Steroid-induced diabetes: Diabetes is a condition in which your body does not produce enough insulin to maintain a normal blood glucose (sugar) level. Long-term use of corticosteroids may cause diabetes, which must be treated in the same way as it is for other people with diabetes.

After a thorough physical and dietary exam, your doctor will probably place you on a special diet. You should consult with a registered dietitian who can help you understand the various aspects of the diet, and learn to plan your meals more easily. You may also have to take a drug to help keep your glucose levels within normal limits. For some people, a pill may be prescribed; for others, insulin given by injection may be necessary.

If you are diagnosed with steroid-induced diabetes, ask your doctor or nurse to refer you to a diabetes education program. These programs help newly diagnosed people with diabetes learn about their disease and manage their condition so that they continue to live a healthy and productive life. If a program is not available where you live, a registered dietitian should be able to give you the information you need.

Kidney disease: Because the kidney is often affected by lupus, your doctor will probably order a variety of tests every so often to see how well your kidneys are working. If your doctor determines that your lupus has affected your kidneys, the goals for treating the problem will be to preserve as much kidney function as possible and prevent the condition from getting worse. Along with other treatment options, you may be placed on a low-sodium (salt), low-potassium, or low-protein diet. A registered dietitian can help you plan meals for these diets.

Cardiovascular disease: Cardiovascular complications of lupus include atherosclerosis and high blood pressure. Atherosclerosis is a condition in which fatty deposits build up on the inside of the arteries. These deposits can reduce or block blood flow. High blood pressure increases the risk of having a heart attack or stroke. High blood pressure can happen when lupus damages the kidneys, which help regulate blood pressure.

If your doctor determines that you have risk factors for atherosclerosis, you will probably be placed on a low-fat diet and an exercise plan. These will help you lower your blood cholesterol level and maintain a good body weight. If you have high blood pressure, you may be placed on a low-sodium diet, medication, or both. These will help reduce your blood pressure to within normal limits.

Patient Information Sheet

Sexuality and Lupus

The constant pain and fatigue associated with lupus may make it difficult to cope with the physical and emotional aspects of sex. In addition, some medications used to treat lupus can cause problems. Some of these drugs may decrease your sex drive. Other drugs may lessen sexual arousal or make it difficult to achieve an orgasm.

Some people with lupus also have a condition known as Raynaud's phenomenon. Exposure to cold causes spasms in the small blood vessels of the finger and toes. This reduces blood flow and may cause fingers and toes to turn white or blue and numb. During sex, the flow of blood increases to the genital area and decreases to other areas of the body, including the fingers. This can cause the numbness and pain of Raynaud's phenomenon to occur.

Other problems also can interfere with sexual activity, such as oral and genital sores, vaginal dryness, and yeast infections. You may feel less attractive because of skin rashes that are difficult to control.

Your partner may not understand the changes in your desire, the fact that you may feel unattractive, or the physical problems you are experiencing. He or she may think you are no longer attracted to him or her. On the other hand, you may feel your partner is avoiding you, when he or she is trying to be sensitive to your needs and is afraid of hurting you or causing you more pain during sexual contact.

These issues may be hard for you to talk about. However, a mutual willingness to have open and honest discussions with your partner can play an important part in understanding the issues that are affecting your relationship. If the two of you cannot resolve your problems together, seek help from your doctor, nurse, or a counselor experienced in working with people who have lupus.

ADDITIONAL NOTES

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CARING FOR YOURSELF

- Keep a healthy attitude about yourself. Being positive can play an important part in maintaining your sexuality.
- If you notice a change in sexual desire after starting a new medication, tell your doctor or nurse.
- Ask your doctor if he or she can prescribe an anti-inflammatory or pain medication that you can take before having sex.
- Be sure you are well rested. Consider taking a nap just before sexual activity.
- Relax and ease some of the pain with a warm shower or bath just before sexual activity.
- If you have Raynaud's phenomenon, increase circulation to your fingers and toes by taking a warm bath before sex. Raising the temperature in the bedroom will also help.
- If you have vaginal dryness, use a water-based personal lubricant during sex.
- If you have a vaginal yeast infection, call your doctor so that he or she can prescribe the medication you need. Yeast infections are easily treated.
- If some physical problems make certain sexual activity difficult, don't be afraid to explore with your partner other ways to achieve mutual pleasure and satisfaction.

From *LUPUS: A Patient Care Guide for Nurses and Other Health Professionals*
National Institute of Arthritis and Musculoskeletal and Skin Diseases/National Institutes of Health

Pregnancy and Lupus

Twenty years ago, medical textbooks said that women with lupus should not get pregnant because of the risks to both the mother and unborn child. Today, most women with lupus can safely become pregnant. With proper medical care you can decrease the risks associated with pregnancy and deliver a normal, healthy baby.

To increase the chances of a happy outcome, however, you must carefully plan your pregnancy. Your disease should be under control or in remission before conception takes place. Getting pregnant when your disease is active could result in a miscarriage, a stillbirth, or serious complications for you. It is extremely important that your pregnancy be monitored by an obstetrician who is experienced in managing high-risk pregnancies and who can work closely with your primary doctor. Delivery should be planned at a hospital that can manage a high-risk patient and provide the specialized care you and your baby will need. Be aware that a vaginal birth may not be possible. Very premature babies, babies showing signs of stress, and babies of mothers who are very ill will probably be delivered by cesarean section.

One problem that can affect a pregnant woman is the development of a lupus flare. In general, flares are not caused by pregnancy. Flares that do develop often occur during the first or second trimester or during the first few months following delivery. Most flares are mild and easily treated with small doses of corticosteroids.

Another complication is pregnancy-induced hypertension. If you develop this serious condition, you will experience a sudden increase in blood pressure, protein in the urine, or both. Pregnancy-induced hypertension is a serious condition that requires immediate treatment, usually including delivery of the infant.

The most important question that pregnant lupus patients ask is, "Will my baby be okay?" In most cases, the answer is yes. Babies born to women with lupus have no greater chance of birth defects or mental retardation than do babies born to women without lupus. As your pregnancy progresses, the doctor will regularly check the baby's heartbeat and growth with sonograms. About 25% of lupus pregnancies end in unexpected miscarriages or stillbirths. Another 25% may result in premature birth of the infant. Although prematurity presents a danger to the baby, most problems can

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CARING FOR YOURSELF

- Keep all of your appointments with your primary doctor and your obstetrician.
- Get enough rest. Plan for a good night's sleep and rest periods throughout the day.
- Eat a sensible, well-balanced diet. Avoid excessive weight gain. Have your obstetrician refer you to a registered dietitian if necessary.
- Take your medications as prescribed. Your doctor may have you stop some medications and start or continue others.
- Don't smoke, and don't drink alcoholic beverages.
- Be sure your doctor or nurse reviews with you the normal body changes that occur during pregnancy. Some of these changes may be similar to those that occur with a lupus flare. Although it is up to the doctor to determine whether the changes are normal or represent the development of a flare, you must be familiar with them so that you can report them as soon as they occur.
- If you are not sure about a problem or begin to notice a change in the way you feel, talk to your doctor right away.
- Ask your doctor or nurse about participating in childbirth preparation and parenting classes. Although you have lupus, you have the same needs as any other new mother-to-be.

be successfully treated in a hospital that specializes in caring for premature newborns.

About 3% of babies born to mothers with lupus will have neonatal lupus. This lupus consists of a temporary rash and abnormal blood counts. Neonatal lupus usually disappears by the time the infant is 3–6 months old and does not recur. About one-half of babies with neonatal lupus are born with a heart condition. This condition is permanent, but it can be treated with a pacemaker.

PLANNING YOUR PREGNANCY

You and your spouse or partner should talk to your doctor about the possibility of pregnancy. You and the doctor should be satisfied that your lupus condition is under good control or in remission. Your doctor should also review potential problems or complications that could arise during the pregnancy, their treatment, and outcomes for both you and the unborn child.

You should select an obstetrician who has experience in managing high-risk pregnancies. Additional experience in managing women with lupus is also good. The obstetrician should be associated with a hospital that specializes in high-risk deliveries and has the facilities to care for newborns with special needs. It is a good idea to meet with the obstetrician before you become pregnant so that he or she has an opportunity to evaluate your overall condition before conception. This meeting also will give you the opportunity to decide if this obstetrician is right for you.

Check your health insurance plan. Make sure that it covers your health care needs and those of the baby and any problems that may arise.

Review your work and activities schedule. Be prepared to make changes if you are not feeling well or need more rest.

Consider your financial status. If you work outside the home, your pregnancy and motherhood could affect your ability to work.

Develop a plan for help at home during the pregnancy and after the baby is born. Motherhood can be overwhelming and tiring, and even more so for a woman with lupus. Although most women with lupus do well, some may become ill and find it difficult to care for their child.

ADDITIONAL NOTES

Continued on next page

AFTER THE BABY IS BORN (THE POSTPARTUM PERIOD)

Be sure your doctor or nurse reviews with you the physical and emotional changes that occur as your body returns to normal. These changes are the same as those experienced by women who do not have lupus.

Be aware that postpartum complications can arise. In addition to those that can occur to any woman who has been pregnant, you might develop a lupus flare.

Try to breastfeed your baby. It is the ideal, low-cost way to provide nutrition for your baby in the first weeks or months of life. It takes time for mothers and babies to learn how to breastfeed and it may take a few weeks to get adjusted. Because breastfeeding can sometimes be a challenge, ask your doctor or nurse for help so you do not become discouraged. Sometimes, though, breastfeeding may not be possible for the following reasons:

- A premature baby may not be able to suck adequately. Feeding your baby through a tube at first and then by bottle may be necessary. However, you may still be able to pump your breast milk for your baby.
- If you are taking corticosteroids, you may not be able to produce enough milk.
- Some medications can pass through your breast milk to your infant. It will be up to your doctor to decide if breastfeeding is safe if you are taking any of these medications.
- Because breastfed infants tend to eat more frequently than do bottle-fed infants, breastfeeding can be very tiring. You may want to switch to a bottle and formula if breastfeeding becomes too tiring.

Be confident, though, that whichever method you choose to use to feed your baby, it will be the right decision for everyone concerned.

Before you leave the hospital, discuss birth control options with your doctor. Because it would be unwise for you to become pregnant again soon after giving birth, be sure to use an effective birth control method. REMEMBER: *You can get pregnant before your period begins again; also, breastfeeding and withdrawal of the penis before ejaculation are not effective birth control methods.*

ADDITIONAL NOTES

From *LUPUS: A Patient Care Guide for Nurses and Other Health Professionals*
National Institute of Arthritis and Musculoskeletal and Skin Diseases/National Institutes of Health

Patient Information Sheet

Nonsteroidal Anti-Inflammatory Drugs (NSAIDs)

NSAIDs are often used to reduce pain and inflammation in patients who have mild systemic lupus erythematosus (SLE). Many different types of NSAIDs exist, some of which you can buy without a doctor's prescription. These are called "over-the-counter" drugs. Examples of over-the-counter NSAIDs include aspirin, Motrin,[1] Orudis, and Anaprox. Tylenol is not an NSAID and is not used to reduce the inflammation of lupus.

Although all NSAIDs appear to work in the same way, there are differences among them. Not every NSAID has the same effect on every person. Also, you may find that one NSAID works well for a while, then for some unknown reason, it doesn't work well any more. Your doctor will probably switch you to a different NSAID to get the same helpful effects you had with the first one.

INSTRUCTIONS

The brand name of your NSAID is: _____

The strength or dose of the NSAID ordered for you is: _____

Take the NSAID _____ time(s) per day.

The best time(s) to take your NSAID: _____

Additional instructions: _____

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[1] Brand names included here and in this book are provided as examples only, and their inclusion does not mean that these products are endorsed by the National Institutes of Health or any other Government agency. Also, if a particular brand name is not mentioned, this does not mean or imply that the product is unsatisfactory.

POSSIBLE SIDE EFFECTS

These include upset stomach, headache, ringing in the ears, dizziness, rash, itching, easy bruising, fluid retention, blood in the stool, and _____

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PRECAUTIONS

You may use NSAIDs cautiously during pregnancy, but do not take them during the first 3 months of your pregnancy or just before delivery. NSAIDs appear in breast milk and should be used cautiously if you are breastfeeding.

Some patients taking NSAIDs become more sensitive to sunlight. Use sunblock and protective clothing; avoid exposure to sunlight.

Do not take more than the recommended dose.

Do not take NSAIDs with other drugs, including over-the-counter medications, without first checking with your nurse or doctor. Over-the-counter medications are medications that you can buy without a doctor's prescription.

Tell any nurse, doctor, or dentist who is taking care of you that you are taking NSAIDs for your lupus.

Since NSAIDs can cause stomach and intestinal upset and irritation, take them with food or after meals. You should also avoid alcoholic beverages, because alcohol can aggravate these stomach and intestinal problems. Check with your doctor for guidance on these issues.

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Patient Information Sheet

Antimalarials

Antimalarials are very effective in controlling lupus arthritis, skin rashes, mouth ulcers, and other symptoms such as fatigue and fever. They are used to manage less serious forms of systemic lupus erythematosus (SLE) in which no organs have been damaged. Antimalarials are also very effective in the treatment of discoid lupus erythematosus (DLE).

Although antimalarials may be very effective in controlling your lupus, their use takes patience. It may take weeks or months before you see any change in symptoms from the use of these drugs.

INSTRUCTIONS

The brand name of your antimalarial is: _____

The strength or dose of the antimalarial ordered for you is: _____

Take the antimalarial _____ time(s) per day.

The best time(s) to take your antimalarial: _____

Additional instructions: _____

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POSSIBLE SIDE EFFECTS

These include stomach upset, loss of appetite, vomiting, diarrhea, blurred vision, difficulty in focusing, headache, nervousness, irritability, dizziness, muscle weakness, dry and itchy skin, mild hair loss, rash, change in skin color, unusual bleeding or bruising, and _____

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PRECAUTIONS

There is a small chance that antimalarials will harm a fetus. If you are considering pregnancy, your doctor may take you off the drug.

Do not take more than the recommended dose.

Do not take this drug with other drugs, including over-the-counter medications, without first checking with your nurse or doctor. Over-the-counter medications are medications that you can buy without a doctor's prescription.

Tell any nurse, doctor, or dentist who is taking care of you that you are taking an antimalarial for your lupus.

WARNING!

A possible, serious side effect of this drug is damage to the retina of the eye. Although this is rare with the low doses of drug that are prescribed, it is extremely important that you have a thorough eye examination before starting treatment with this drug and every 6 months after that.

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Patient Information Sheet

Corticosteroids

Corticosteroids are very powerful drugs that reduce inflammation in various tissues of the body. These drugs are used to treat many of the symptoms of lupus that result from inflammation. You can take this medication as pills or by injection. Corticosteroid creams or ointments are also available to treat skin rashes caused by lupus. Most lupus symptoms respond quickly to corticosteroids. Prednisone is a corticosteroid that is often used to treat lupus.

The decision to begin corticosteroids is a big one and depends on your needs. Some patients may need to take the drug for a short time only, until disease symptoms get better or go away. Others with more serious or life-threatening problems may require higher doses of the drug for longer periods of time. In general, once your lupus symptoms have responded to treatment, you will gradually take less and less of the drug until you can stop completely. If it is not possible for you to stop the drug completely, your doctor will give you the smallest amount possible to keep symptoms under control.

Doctors are careful about prescribing corticosteroids because many complications are associated with taking this drug. As a result, it is important to take the drug exactly as prescribed. People who have been taking corticosteroids for a long time may need higher doses of the drug before, during, or after a physically stressful event, such as surgery.

INSTRUCTIONS

The brand name of your corticosteroid is: _____

The dose of the corticosteroid that is ordered for you is: _____

Take the corticosteroid _____ time(s) a day.

The best time(s) to take your corticosteroid: _____

You may take your corticosteroid with food or after meals if stomach upset occurs.

POSSIBLE SIDE EFFECTS

These include changes in appearance (such as acne or increased facial hair); development of a round or moon-shaped face; thin, fragile skin that bruises easily; or movement of body fat to the trunk. You might also experience mood changes, personality changes, irritability, agitation, or depression.

Other possible side effects include increased appetite and weight gain, poor wound healing, headache, glaucoma, irregular menstrual periods, peptic ulcer, muscle weakness, osteoporosis, steroid-induced diabetes, and osteonecrosis (damage to the hip joint that leads to severe arthritis).

PRECAUTIONS

Because corticosteroids cross the placenta, they are used cautiously during pregnancy. The drugs appear in breast milk, so if you are taking large doses, you should not breastfeed.

Avoid exposure to infections. Stay away from crowds and people known to have colds, the flu, or other infections.

Schedule regular vision checkups and report any problems with your vision to your doctor or nurse.

Talk with a registered dietitian to find out how to prevent excess weight gain and minimize certain drug effects on the body.

Do not take this drug with other drugs, including over-the-counter medications, without first checking with your nurse or doctor. Over-the-counter

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Additional instructions: _____

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medications are medications that you can get without a doctor's prescription.

Tell any nurse, doctor, or dentist who is taking care of you that you are taking corticosteroids for your lupus.

WARNINGS!

Do not take this drug if you have ever had an allergic reaction to it.

Carry medical identification and wear a bracelet to alert medical personnel that you take a corticosteroid.

NEVER MISS A DOSE. Take this drug exactly as ordered.

IF YOU DO MISS A DOSE, call your nurse or doctor immediately to find out when you should take the missed dose.

NEVER STOP THE MEDICATION SUDDENLY. Your adrenal glands, which are located just above your kidneys, normally make corticosteroids in small amounts. These corticosteroids are important for many body functions. When you take corticosteroid medication, your body begins to make much less than usual, or even stops completely. If you suddenly stop taking your medication, you may have a problem because your adrenal glands won't have had time to make the corticosteroids you need. This problem is called "adrenal insufficiency."

Signs of adrenal insufficiency include weakness, fatigue, fever, weight loss, vomiting, diarrhea, and abdominal pain. If you experience any of these problems, call your nurse or doctor immediately.

From *LUPUS: A Patient Care Guide for Nurses and Other Health Professionals*
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Patient Information Sheet

Azathioprine

Azathioprine is a drug that acts to suppress the work of the immune system. It is used mainly in organ transplantation to prevent the body from rejecting the new organ. The drug is also used in patients with lupus who have damage to their kidneys or other organs, muscle inflammation, or advanced arthritis. Azathioprine helps to reduce symptoms and damage to the affected organs. It can also help achieve a remission of the disease.

Another benefit of azathioprine is that it reduces or even eliminates the need for corticosteroid therapy. This means that patients do not have to have the unpleasant side effects that occur with corticosteroids. Immunosuppressives like azathioprine, however, can have their own serious side effects. Your doctor must work closely with you to make sure that the amount of the drug you are taking gives you the benefits you need with as few side effects as possible.

INSTRUCTIONS

The brand name of your azathioprine is: _____

The strength or dose of the azathioprine ordered for you is: _____

Take the azathioprine _____ time(s) per day.

The best time(s) to take your azathioprine: _____

You may take azathioprine with food if stomach upset occurs.

Additional instructions: _____

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POSSIBLE SIDE EFFECTS

These include stomach upset, nausea, vomiting, abdominal pain, mouth ulcers, darkened urine, pale stools, jaundice (yellowing of the skin or white portion of the eyes), unusual bleeding or bruising, signs of infection (such as chills, fever, sore throat, or fatigue), and

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PRECAUTIONS

Do not take more than the recommended dose.

Avoid exposure to infections. Stay away from crowds and people known to have colds, the flu, or other infections.

Do not take this drug with other drugs, including over-the-counter medications, without first checking with your nurse or doctor. Over-the-counter medications are medications that you can buy without a doctor's prescription.

Tell your nurses, doctors, or dentist that you are taking azathioprine for your lupus.

WARNING!

Use of this drug presents a definite risk to the fetus. Use an effective birth control measure during treatment and for 12 weeks after ending treatment. Azathioprine may pass into breast milk, so consult your doctor before breastfeeding.

From *LUPUS: A Patient Care Guide for Nurses and Other Health Professionals*
National Institute of Arthritis and Musculoskeletal and Skin Diseases/National Institutes of Health

Patient Information Sheet

Cyclophosphamide

Cyclophosphamide is a drug used to treat a number of cancers, and it is used to treat patients with lupus when major organs, such as the kidneys, are affected. It is also used to treat severe inflammation that has not responded to corticosteroids. In lupus, the immune system is too active. Cyclophosphamide slows down the immune system so that disease activity can be reduced.

Cyclophosphamide is a very powerful drug. It can have a number of effects on the body. As a result, it is important that you understand how it is used to treat your lupus. You will need to work closely with your doctor and nurse to make sure that the amount of the drug you are taking gives you the benefits you need with as few side effects as possible.

INSTRUCTIONS

The brand name of your cyclophosphamide is: _____

The strength or dose of the cyclophosphamide ordered for you is: ____

Take the cyclophosphamide _____ time(s) per day.

The best time(s) to take your cyclophosphamide: _____

Drink at least 2 quarts of water every day while taking this drug. That is equal to four big (16-oz.) glasses.

Additional instructions:

_____

_____

_____

_____

_____

POSSIBLE SIDE EFFECTS

These include nausea, vomiting, loss of appetite, mouth ulcers, fatigue, temporary hair loss, unusual bleeding or blood in the urine, shortness of breath, loss of menstrual periods, impotence, sterility, or signs of infection (such as increased temperature, sore throat, or flu symptoms).

Tell your nurse or doctor right away if you have any side effects.

PRECAUTIONS

Do not take more than the recommended dose.

Avoid exposure to infections. Stay away from crowds and people known to have colds, the flu, or other infections.

Tell any nurse, doctor, or dentist who is taking care of you that you are taking cyclophosphamide for your lupus.

WARNING!

Do not take this drug if you suspect you are pregnant. Cyclophosphamide causes birth defects. You must use an effective birth control method while you are taking this medication. You should consider pregnancy only after treatment has been stopped and your doctor says you are healthy enough to become pregnant.

Long-term therapy with cyclophosphamide may leave a woman unable to produce eggs, or a man unable to produce sperm. This means permanent sterility. If you want to have a baby in the future, talk to your doctor about the option of storing your eggs or sperm before beginning therapy.

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The mission of the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS), a part of the Department of Health and Human Services' National Institutes of Health (NIH), is to support research into the causes, treatment, and prevention of arthritis and musculoskeletal and skin diseases, the training of basic and clinical scientists to carry out this research, and the dissemination of information on research progress in these diseases. The National Institute of Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse is a public service sponsored by the NIAMS that provides health information and information sources. Additional information can be found on the NIAMS Web site at www.niams.nih.gov.

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